ADULT CONGENITAL HEART DISEASE: ASSESSING BARRIERS TO CARE IN SASKATCHEWAN

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Abstract:

Background: The majority of patients born with congenital heart disease (CHD) now live to adulthood due to advanced surgical techniques and pediatric cardiology expertise. Facilitating successful transition to adult care has been deemed a priority. Sufficient capacity of adult CHD providers to handle the increased number of patients, improved communication between pediatric providers and their patients regarding transition, and health care autonomy have been identified as a predictors of successful transition.

Methods: This project assessed transition success and adherence to follow-up recommendations among ACHD patients in Saskatchewan, Canada. We contacted young adult patients with CHD who were deemed to require lifelong cardiac care and should have transitioned to adult care between 2007 and 2014. We completed a telephone survey to assess demographics, socioeconomic factors, and care preferences.

Results: Of the 106 patients contacted, 32 consented to participate in the telephone survey portion of the study (30% response rate). Nineteen of the respondents (59%) were male with an average age of 22 +/- 2 years. All but one of the participants, who immigrated from the Phillipines at age 14, were born in Canada and their first language was English. Twenty-two respondents (69%) had CHD of moderate to severe complexity (n=22, 69%) while the remainder (n=10, 31%) had simple CHD. Knowledge of congenital heart disease was low with only 63% of

participants able to describe their congenital heart condition. Only 69% were receiving guideline-based follow-up for their ACHD although almost all (91%) were being followed by a cardiologist. Seventy-five per cent of participants (n=24) received a low overall score on the Krantz Health Opinion Survey (6 or less), indicating a lack of health care autonomy.

Conclusions: Young adults in Saskatchewan have poor knowledge of their underlying congenital heart disease. They are not receiving guideline-based care for their ACHD. ACHD patients also express passive preferences toward receiving information about their health and decision-making. Further research should focus on behavioural interventions to improve self-care practices, while investigating and acknowledging the psychological effects of chronic illness on young adults.

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1.0 Introduction

1.1 Statement of the Problem

Modern surgical techniques to repair congenital heart defects have dramatically increased the survival rate of children born with heart disease. About 90% of these children are now living into adulthood.[1] The disease is never cured, only repaired, so they require specialized care throughout their lives to navigate pregnancy, late surgical complications, and the additive effects of acquired adult diseases. In 2000, there were an estimated 800,000 adult congenital heart patients living in the US.[2] The ACC (American College of Cardiology) estimates that, in the next decade, 1 in 150 adults in the developed world will be living with some form of congenital heart defect, from mild to complex lesions.[3] Based on current population estimates from Quebec (the only Canadian data available), at least 150,000 Canadians are living with ACHD with 4,000 to 5,000 in the province of Saskatchewan. Of the patients in Saskatchewan, between 400 and 500 will have complex congenital heart disease[2].

This presents a challenge for adult cardiologists as a new patient population is quickly forming. As children transition from pediatric cardiology into adult paradigms of care, there are significant barriers to continued access. The well-established programs including nurse educators, psychologists, and social workers that are available to pediatric cardiology patients and their families are often inaccessible to adult patients. A survey of US pediatric cardiologists revealed that

the majority (79%) is still providing care to adult patients and a major factor was a lack of adult care provider available.[4] As the number of ACHD patients continues to rise, this approach will no longer be feasible.

In Saskatoon, ACHD patients are followed by cardiologists with no formal ACHD training. There is no nursing or clerical support, and there is no electronic database attaching patients to cardiac diagnoses. The closest pediatric surgery program is in Edmonton, Alberta, which is more than 500km away. Despite evidence that ACHD patients have significant psychosocial challenges, there is no social worker or psychologist affiliated with the Saskatoon ACHD clinic.[5] In Regina, there is one cardiologist with formal training and board certification in ACHD who runs a formal ACHD clinic with nursing and clerical support only. Therefore, in the province of Saskatchewan, there is only one cardiologist with the appropriate training necessary to care for ACHD patients.

1.2 Purpose of the Study

Study of the ACHD population in Saskatchewan has never been undertaken prior to this study. Based on Canadian demographics and discussion with pediatric cardiologists, we know that this population is growing.[2] The purpose of this study was to assess the transition needs and barriers to care of pediatric cardiology patients as they transition to an adult care paradigm. This research will provide a framework for program planning and optimization of the adult care paradigm for ACHD patients in Saskatchewan.

1.3 Theoretical Perspective

This study was undertaken using a deductive reasoning approach attempting to identify barriers to care for ACHD patients. We investigated, in a retrospective manner, the independent factors associated with the dependent variable (loss to follow-up or missed cardiology appointments as an adult).

1.4 Research Questions & Hypothesis

Our hypothesis was as follows: Patients with ACHD face many barriers to transition from a pediatric to an adult care paradigm. These barriers are correlated with loss-to-follow-up from appropriate adult cardiac care.

The barriers we proposed to be associated with loss-to-follow-up included: patient demographics such as sex, age, and complexity of disease; health care system barriers such as lack of adult providers or inadequate knowledge; socioeconomic barriers including involvement of parent/support person or financial constraints; and psychological barriers like anxiety and/or depression (Figure 1.1).

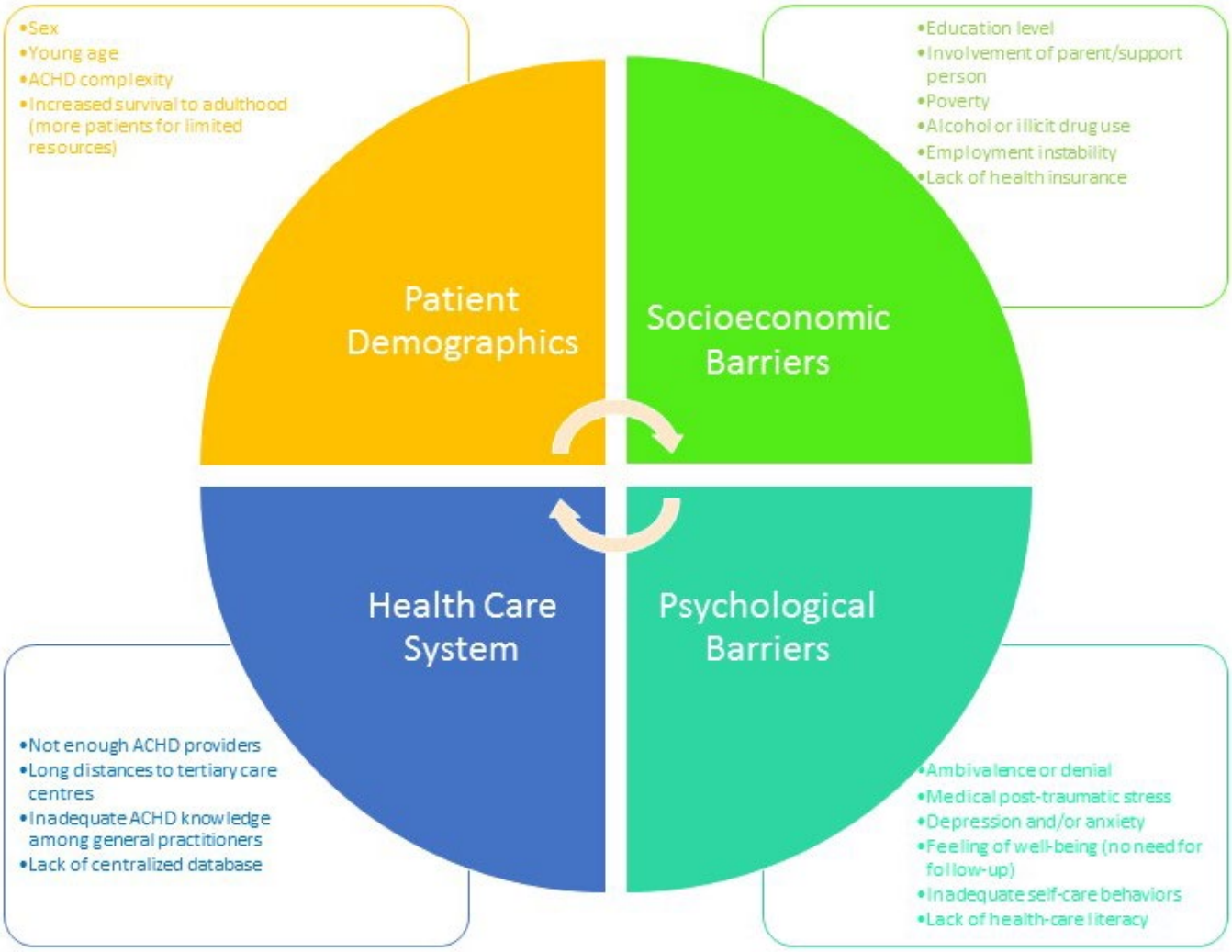


Figure 1.1. Barriers to successful transition from pediatric cardiology care to long-term adult ACHD care.

1.5 Definition of Terms

Transition – The physical and psychological move from pediatric care to adult care paradigms requiring the self-efficacy necessary to take responsibility for one's own health care.

Adult Congenital Heart Disease – A cardiac condition present during childhood (structural or acquired infectious (rheumatic fever valvulopathy)) requiring lifelong follow-up and specialist care in a tertiary care centre.

Loss-to-follow-up – Loss from appropriate specialist care for more than two years from last expected date of follow-up.

1.6 Delimitations and Limitations

Because all pediatric cardiology care in Saskatchewan is centralized in Saskatoon, we were able to identify all patients who graduated from pediatric care in the province in a 7-year time span (2007-2014). A complicating factor was the adult congenital care situation in the province. There is one ACHD cardiologist in Regina and several general cardiologists in Saskatoon who follow transitioned patients and no centralized database. For that reason, it was difficult to access adult records for each patient as they were spread out through many different physician

offices with no electronic medical record. Also, some patients may have moved out of province and are not followed in Saskatchewan any longer.

For that reason, instead of performing a simple chart review of adult patients, a telephone survey was designed to contact transitioned patients. In this manner, we could not verify the responses of the participants regarding their follow-up frequency but we obtained additional information by speaking directly with the patients. One of the limitations was that we only surveyed patients we were able to contact using their last pediatric point of contact. For this reason, we often contacted parents of transitioned children to obtain their most recent contact information. Therfore, we were not able to reach each transitioned patient and the generalizability of our study is subsequently limited.

Another limitation was the retrospective nature of the research and the absence of prospective study. It would be informative to follow patients forward to see whether or not loss-to-follow-up correlates with medical outcome (morbidity/mortality). This study did not provide this type of information but it may set the stage for future research regarding the consequences of loss-to-follow-up.

2.0 Literature Review

2.1 The Demographics of ACHD

The global birth incidence of ACHD, since 1995, is estimated at 9 per 1,000 live births.[6] This amounts to approximately 1.35 million newborns per year worldwide, with the majority of lesions being simple structural abnormalities (ventricular septal defect (VSD), atrial septal defect (ASD), and patent ductus arteriosus (PDA)). Africa has the lowest reported birth rate but this is likely due to underreporting resulting from a lack of health care services and equipment (ie. babies being born in rural areas and dying without a clear diagnosis). Therefore, the global incidence is probably slightly higher than reported.[6] Maternal diabetes, febrile illness or infection, drug exposure, and organic solvent exposure are all known risk factors for giving birth to a child with congenital heart disease.[6] In the last half century, medical advances such as cardiopulmonary bypass machines, critical care units, and modern surgical techniques have significantly improved outcomes for these infants. Without intervention, 30% of them would die within the first year of life.[7] Today, in the developed world, more than 90% of these infants live into adulthood.[1] Deaths in low-middle income countries could be reduced by a further 67% if pediatric surgical programs were widely available.[8]

Using provincial database information from Quebec, the Canadian prevalence of all congenital heart disease in 2007 was estimated at 4 per 1000 adults and the prevalence of severe lesions is 0.38 per 1000 adults.[2] This number is lower than the

estimated 9/1000 birth incidence due to local maternal factors; reduced febrile illnesses, folic acid deficiencies, solvent exposure, and untreated maternal diabetes in Canada. Despite improvements in pediatric surgical techniques, there is also a temporal lag in survival reducing the number of contemporary adults who have survived to adulthood. Also, although Quebec has a provincial database in which all patients are included, it relies upon International Statistical Classification of Diseases and Related Health Problems (ICD) coding which can be problematic in congenital heart disease as adult patients may be coded inaccurately and the true prevalence may be underestimated.[2] The Quebec population also has a unique genetic makeup which makes national generalizability more difficult.

As more patients with severe congenital heart disease survive pediatric surgeries, the adult prevalence is expected to increase. A Belgian study compared patients born between 1990 and 1992 to an anatomically similar cohort born in the early '70s. They found that more than 88% of those born in the early '90s survived to adulthood, a proportion significantly higher than those born in the prior two decades.[1] From 1987 to 2005, overall mortality from congenital heart disease declined by 31% and the median age at death increased by 15 years.[9] Because of the shift in mortality from pediatric life to adulthood, there are currently more adults living with congenital heart disease than children. Overall, 66% of patients with CHD were adults by 2010.[10]

There are over 100 pediatric cardiologists registered with the Canadian Pediatric Cardiology Association but far fewer adult cardiologists trained to care for these patients once they reach adulthood. The incredible success of the last 50 years

has left a gaping hole in resources for these adult patients who now expect and deserve lifelong specialized care. Resource utilization studies have shown that ACHD patients require significantly more health care resources than people of a similar age. Over a five-year period in Quebec, 68% of patients with ACHD visited the ER at least once and more than half of them were hospitalized. These rates are twice as high as those among the general Quebec population. [11] A Dutch study found that patients with ACHD are admitted to hospital with heart failure at significantly higher rates than age-matched peers (1.2 per 1000 patient years vs. 0.1 per 1000 patient years). Median age at first heart failure admission was 46 and the risk of death for an ACHD patient with heart failure is increased five-fold.[12] This indicates that, beyond the simple increase in the number of ACHD patients accessing the health care system, the disease burden of this patient population makes their need of services greater than age-matched peers.

In summary, the number of adults with congenital heart disease is increasing rapidly because of advances in pediatric care. Adult resources have failed to keep pace with the burgeoning patient population.

2.2 Barriers to Care in ACHD

Many adult patients with congenital heart disease are lost to appropriate specialist follow-up. A U.K study examined those with a specific diagnosis requiring lifelong specialized care and frequent adult interventions, Tetralogy of Fallot, and found that 24% of patients had not been seen in a specialist ACHD clinic for more

than 3 years. Most patients were lost for more than 22 years.[13] When patients were contacted by telephone to ascertain why they were lost from care, several felt it was the duty of the health care system to follow-up with them and ensure they were receiving adequate care. Others expressed feelings of ambivalence or denial about their disease. When the charts of lost patients were reviewed, several deaths from cardiac causes may have been preventable.[13]

A period of critical importance in lifelong ACHD care is the transition period. This is the period during which adolescent patients move from the pediatric to adult care paradigm and assume responsibility for their care from their parents and pediatric providers. A Canadian study evaluating patients who transitioned in Toronto in 1980 found that only 47% of patients successfully transferred to adult care.[14] Correlates of successful transfer were: an increased number of pediatric surgeries (indicating increased complexity of congenital heart disease), older age at last pediatric appointment, documentation of a transition discussion in the patient's chart, lack of alcohol or drug use, and attending pediatric appointments independently without a parent.[14]

A recent U.S. study, which is confounded by insurance status in a country without universal health care, showed that only 68% of patients transitioned between 2008 and 2011 made a successful transfer. Those with increased complexity of CHD and those taking medications were more likely to make the move. They found an average of eight missed or rescheduled appointments per patient during the transition period.[15] A Belgian study published in 2011 found that 7.3% of ACHD patients were not receiving any follow-up. Their study confirmed that

male patients with simple lesions and no prior heart surgery were more likely to have lapses in care. Their lost-to-follow-up rate was much lower than previously published studies but they excluded all patients they could not locate and those who did not respond to a mailed survey, therefore, they likely deflated the true rate of loss.[16] A Quebec study revealed similar results regarding correlates of loss-to-follow-up, although they found a higher rate of patients who were not receiving care in an adult cardiology clinic (39% of ACHD patients aged 18-22).[17]

Lack of appropriate transition can place an unnecessary burden on the health care system. A U.S. study found that admissions to hospital following emergency room (ER) visits were twice as high for ACHD patients aged 21-23 as those aged 15-17.[18] Some of these ER visits were likely appropriate but the drastic increase could represent a failure to adequately transition and lack of preventive outpatient care. A U.K. study found that 38% of living adult patients had no follow-up and that, of patients who had died, 48% were not being followed. This suggests that loss to specialist follow-up also correlates with increased mortality from ACHD.[19] A Quebec demographic study linking health care utilization data with diagnostic codes also found a slow increase in referrals to ACHD referral centres which was significantly correlated to a decline in mortality.[20]

Overall, many patients with ACHD are not receiving appropriate specialist follow-up and the reasons are multifactorial. A large multicenter North American study found that, of patients surveyed in an ACHD clinic setting, 42% reported at least a three year gap in follow-up at some point in their adult lives. They found a sensation of well-being, patients who only felt they needed to see a doctor if they felt

unwell, and a lack of education regarding the long-term complications of ACHD were barriers to continuation of specialist care.[21]

A study of outpatient clinic visits at a tertiary care centre in the United Kingdom found that clinical workload for ACHD cardiologists more than tripled from 1990 to 2010.[22] They found that younger patients with indices of socioeconomic deprivation were less likely to attend scheduled clinic visits although gender, ACHD complexity, and travel distance were not predictors of non-adherence. Clinical non-adherence was found to be associated with mortality.[22]

A study in the United States also found increased morbidity in ACHD patients who were lost to follow-up.[23] Those with lapses in care of more than two years in duration were three times more likely to require urgent interventions than their adherent counterparts.[23] Patients lost to care reported being told that there was no need for long-term follow-up and those living independently from their parents were far more likely to have lapses in care.[23] As this was an American study, the authors proposed that many patients were unable to attend clinic visits because of a lack of health insurance.[23]

Other sociodemographic and psychological factors associated with adherence to ACHD care have been identified. Less than half of ACHD patients report adequate self-care behaviours; physical activity, contraception and pregnancy-planning, and monitoring and management of symptoms.[24] Factors associated with improved self-care were older age, male gender, higher levels of education, and adequate health insurance, although a German study found that there was no association between gender and mortality in congenital heart

disease.[25] Depression and poor family support were correlated with lower levels of care. Knowledge about ACHD was not significantly correlated with increased self-care.[24]

It has also been postulated that patients who are "feeling well" do not appreciate the need for long-term follow-up with ACHD professionals. A German study sent a follow-up survey to nearly 2000 patients who were registered as having ACHD and being lost to care for more than five years. The majority of patients were working full-time and 85% of them described themselves as "healthy and fit".[26]

In summary, loss to specialized ACHD care has been associated with both cardiac morbidity and mortality. Factors consistently associated with lapses in care include younger age, a feeling of well-being not requiring follow-up, and less complex ACHD.

2.3 Quality of Care in ACHD

American studies have identified ACHD being considered a pre-existing condition and inability to obtain health insurance as unique factors affecting lifelong care of these patients.[23] Another barrier to care is the simple lack of adult care providers who specialize in congenital heart disease. Canadian research found that only 27 per cent of ACHD patients were followed by cardiologists formally trained in ACHD.[27] The majority of ACHD clinics across the country have dedicated nursing

support (73 per cent) but less than half have clerical support (40 per cent). Only 73 per cent of clinics have electronic ACHD databases.[27]

In Canada and European countries with a single-payer system, there is often a defined age in the second decade at which patients must transition to adult care. In the United States, where many patients are covered through private insurance, the majority of pediatric cardiologists continue to care for adult patients.[28] Pediatric cardiologists cite both emotional attachment to their patients and a lack of appropriately trained adult ACHD cardiologists to whom they can refer as reasons they continue to care for adult patients.[28] This presents a problem as pediatric cardiologists are not trained to care for patients during pregnancy or as they develop concomitant acquired comorbidities. A recent needs assessment in the United States concluded that the current number of trained ACHD physicians and trainees is not nearly sufficient to care for the burgeoning patient population, making the position of pediatric cardiologists even more difficult.[29]

Several studies have examined the quality of care that congenital heart disease patients receive after reaching adulthood. United States, European, Latin American and Canadian guidelines all suggest assessment by a trained ACHD cardiologist in a clinic staffed with specialized nursing support.[3,30,31,32] Experts recommend one ACHD cardiologist for each population of between two and four million people.[31,33] A European survey revealed that only 19% of "specialist centres" complied with an optimal care structure.[34] The authors defined "optimal care" as performing more than 50 congenital heart surgeries per year and involving nurse specialists in the routine care of ACHD patients.[34]

Other areas of quality control research have included pre-pregnancy counseling, pregnancy management, and compliance with medication recommendations for ACHD patients. A U.K study evaluated patients with a specific lesion; aortic coarctation. These patients have congenital heart disease of moderate complexity that is often repaired in early childhood but requires lifelong follow-up with an ACHD specialist.[35] They have a high-risk of developing early-onset hypertension and require close monitoring and treatment during pregnancy.[35] A review at their first visit to a new specialist ACHD center revealed that 52% of women referred had given birth with no cardiology support. More than half of new patients to the clinic were prescribed medications for hypertension at their first visit and almost all required advanced aortic imaging.[35] This indicates that they were not receiving guideline-based care from their general practitioners prior to referral to the specialist centre. A German study examining self-reported contraception and sexual health behaviours found that only 52% of women with ACHD report being counselled about sexual health and pregnancy by a health practitioner and many of these discussions were initiated by the patients themselves rather than a physician.[36]

Despite evidence that global ACHD care is suboptimal, there is evidence that one quality indicator is being met. Several subsets of patients with ACHD should take antibiotics prior to dental procedures to prevent infection on their heart valves.[37] A Canadian study reviewed clinic letters of ACHD patients and found that dental hygiene and appropriate antibiotic prophylaxis were addressed in the majority of patient charts (73 per cent).[37]

Overall, global care of ACHD patients is clearly suboptimal, often as a result of inadequate resources, patient factors, and a lack of expertise in management of these complex patients during adult life.

2.4 Living with ACHD: Quality of Life and Mental Health

As more congenital heart patients transition to adult care, the previous research focus on morbidity/mortality has shifted to allow for more study of the lived experience of ACHD patients and interventions to improve their quality of life. Being born with a congenital heart defect, undergoing childhood surgery, and living with physical limitations in childhood can have long-lasting effects. Frequent hospitalizations and multiple surgeries can interrupt traditional schooling. A French study found that patients with complex congenital heart disease attain lower educational levels than their healthy peers, especially those who had medical complications or more than one surgery in childhood.[38] The same study identified that more than half of patients with complex lesions were unemployed in adulthood and many were inactive as they were told to refrain from physical activity in childhood.[38] A similar Dutch study revealed that men with ACHD were twice as likely to be unemployed than their age-matched peers and women with ACHD were more likely to be engaged in part-time work.[39] Young patients with ACHD (less than age 40) had lower incomes and were less likely to be in a relationship than a reference group of healthy adults.[40] A systematic review of studies assessing standardized quality of life measures found that physical functioning and overall

general health were decreased in patients with moderate-severe cardiac lesions but not in patients with disease graded as simple in complexity.[41]

In addition to the functional challenges associated with living with ACHD, many patients have lingering psychological trauma from childhood exposure to the medical system. Patients with ACHD have higher rates of anxiety and depression than age-matched counterparts.[42] Many patients with ACHD deny or minimize the impact of the illness on their lives and that denial is associated with avoidance of long-term care.[42] Patients with ACHD who endorse depression and anxiety symptoms are more likely to abuse substances (alcohol, cigarettes) which are patently deleterious to their cardiac health and longevity.[43] A North American study identified that more than half of young adults and a quarter of adolescents with ACHD self-report significant substance abuse (binge drinking, cigarette smoking, marijuana or illicit drug use).[44] This rate is similar to that among healthy young adults even though the majority of ACHD patients acknowledge that their substance abuse will increase or hasten complications from their heart disease.[44]

The relationship between ACHD patients and their parents has been evaluated in qualitative studies. Parents of adolescents with congenital heart disease are heavily involved in their medical care (95% attend all doctor visits and 45% administer all of their teenager's medications) and only half felt their child was mature enough to take responsibility for their health care once they reached the age of transition.[45] Parents of ACHD children feel a strong sense of responsibility to obtain the best care available for their child while also providing a normal family life.[46] This can add significant emotional or financial strain to a marriage. A U.S.

demographic study found that parents of children with CHD had between a two and three times higher rate of divorce than women who gave birth to healthy children.[47]

ACHD adults also face significant interpersonal challenges, including feeling different and being isolated socially, which make intimate relationships fraught with complexity.[48] In addition, the risk of transmission of congenital heart disease to offspring coupled with the possibly deleterious cardiac effect of pregnancy on women with ACHD make partner selection and long-term relationships more difficult. A study of sexual health behaviours and reproductive concerns in young adults with congenital heart disease found that fewer adolescents were sexually active when compared to healthy peers but those that were engaged in risky sexual behaviours (multiple partners, no contraception, substance abuse mixed with sexual encounters).[49] Alarmingly, very few women (35% with complex CHD and 5% with moderate CHD) felt that pregnancy would negatively affect their cardiovascular health despite abundant evidence that pregnancy is a significant cardiac stressor that is known to precipitate complications in women with ACHD.[49]

Recently, the phenomenon of medical post-traumatic stress disorder (PTSD) is being recognized. A study in patients with ACHD found that the self-reported rate of PTSD symptoms was significantly higher than in the general population (11-21% vs. 3.5% respectively).[50] Less than half of patients who screened positive for medical PTSD were receiving mental health treatment and less than five per cent had the diagnosis notated in their medical chart.[50] The long-term effects of medical PTSD and its influence on care-seeking behaviour has yet to be studied.

2.5 The Challenge of Transition

The period of transition from pediatric to adult care has been defined as a lost opportunity. Because of differences in intellectual and emotional maturity of adolescents, it is often difficult to assess and individualize the right time for transfer to an adult care paradigm.[51] A successful transition process provides guidance to the adolescent regarding exercise, career planning, contraception, pregnancy, and travel. An abrupt transition can overwhelm both the patient and their family, leading to a lack of follow-up.[51]

Qualitative studies of adolescents undergoing transition have revealed that ACHD teenagers have anxiety about the need to understand their own heart condition and uncertainty about using medical terms.[52] In fact, research has shown that adolescents at the time of transition often have poor understanding of their underlying congenital heart defect.[53] Less than half of Belgian teenagers surveyed at the time of transition could properly name their heart defect and outline possible late complications necessitating follow-up.[53] Teenagers with ACHD also have poor baseline understanding of endocarditis prevention, sexual health, and cardiovascular risk factors.[54]

A similar American study found that only a dismal nine per cent of adolescents could correctly identify all of the conditions of which they are at risk because of their ACHD.[55] A survey conducted in Alberta showed that only 54 per cent knew the medical name of their heart condition, and only 44 per cent could describe their condition in lay language.[56] Parents of children with congenital heart

disease fared better, with 71 per cent able to provide the medical name of their child's condition.[57] ACHD patients also have significant knowledge deficits regarding pregnancy and obtaining health insurance despite their pre-existing condition.[58] They also consistently overestimate their life expectancy contrary to available evidence and believe they will live into their eighth decade similar to their healthy peers.[59] This lack of understanding of anatomy and future complications could lead to a lower level of vigilance regarding symptoms of deterioration.

Narrative analyses of adolescents undergoing the transition process have identified the benefits of allied health care professionals in easing the difficulty of transition from pediatric to adult care paradigms.[60] A formal and transparent framework for successful transition has been suggested as a way to minimize anxiety surrounding the process.[60] ACHD patients describe the transition as a gradual process of taking responsibility for their own health care and not just a physical move from one facility to another. They suggested that knowing well in advance the timeline for transition, having contact and background information about their new adult cardiologist, and allowing their parents to accompany them to their first adult visit were measures to minimize the feelings of uncertainty surrounding transition.[61] Mentorship from older patients with ACHD has been identified by patients as a positive strategy for becoming more educated about their disease and knowing what to expect in the future with 60% of patients indicating they would like to be involved in such a program.[62] Despite high levels of internet and social media use by young adults with ACHD, 55% say they have not accessed information about their heart condition on the internet.[62] This may be a result of

denial and the fear of uncovering negative prognostic information but it may be simply a lack of appropriate resources available. Therefore, mentorship programs and social media may be areas for intervention that could increase transition effectiveness.

Validated questionnaires evaluating self-reported health status found that there was minimal change over time in adolescents during the transition process with the exception of emotional functioning which declined as patients reached young adulthood.[63] This may quantify the stress and uncertainty associated with transition and taking responsibility for one's future health. Overall, the concept of self-efficacy has been identified as a necessary step to healthy transition. Some experts advocate that adolescents should be assessed individually and transition when they have reached a level of maturity at which they are capable of managing their own care.[64] Importantly, some patients have unique neurocognitive disabilities as a result of their congenital heart disease and may never be fully capable of managing their health care independently. For this reason, the "one size fits all" approach to transition is inappropriate and each patient should be assessed in a framework where their particular challenges should be accommodated.[51]

A systematic review of research evaluating transition effectiveness among teenagers with ACHD identified several factors that consistently predicted a successful transition; a belief that lifelong specialized care was necessary, poorer overall health status, attendance at pediatric appointments without a parent, and direct referral from their pediatric cardiologist to an ACHD provider.[65]

Despite a preponderance of evidence indicating that the period of transition is critical in establishing lifelong self care behaviours in ACHD patients, and that a defined transition program is necessary to facilitate success, less than one third of pediatric cardiology programs provide structured transition preparation.[66] A retrospective chart review revealed that only 10 per cent of adolescent patients have documented education regarding pregnancy, long-term symptoms, substance abuse, and education/career planning.[15] Little research has been done to evaluate specific transition programs and quality improvement research regarding ACHD transition is scarce.

One large study evaluating knowledge interventions in transitioning ACHD patients is the Health, Education and Access Research Trial (HEART-ACHD).[67] This was an intervention study where patients were provided with an educational session about their ACHD where they created a health passport with allied health professionals. They completed a survey assessing their knowledge about their ACHD before and after the intervention. There were significant increases in knowledge after the intervention and 60% of patients reported that the health passport was a useful tool.[67] These patients are being followed longitudinally to identify whether or not improvement in ACHD knowledge correlates with lifelong adherence to specialized care and clinical outcomes.

Although there is little research evaluating specific transition programs in adolescents with ACHD, interventions have been evaluated for adolescents living with other chronic illnesses. A systematic review of transition interventions published in 2015 identified four programs for patients with Type I diabetes and

one for adolescents with sickle cell disease.[68] Programs which included a health care navigator, joint visits with pediatric and adult physicians, and tours of adult facilities prior to transition showed significantly higher rates of success.[68] A grounded theory study found that the concept of "health care advocacy" was important for transition of adolescents with chronic disease.[69] The advocate could be a member of the health care team, a family member, or the patient themselves. They found that preparation for taking on an adult role may be more important to successful transition than medical knowledge.[69]

In summary, the 2008 ACC Guidelines for management of patients with congenital heart disease recommend that, to aid in the transition process, patients should have access to a cardiologist and allied healthcare professionals trained in ACHD. They also recommend educational and outreach programs during the transition period, and afterwards, to maintain continuity of care and to recapture patients who may have become lost to follow-up during the transition process.[3]

This study aimed to assess the rate of successful transfer from pediatric cardiology to adult care in Saskatchewan and identify correlates of unsuccessful transfer. In Saskatoon, there are no cardiologists nor allied health professionals formally trained in ACHD and there is no defined transition clinic. Many patients live in rural or remote northern communities, and there is a large proportion of First Nations patients relative to the rest of Canada, making our patient population unique.

3.0: Methods

3.1 Research Design:

This was a retrospective study using a telephone survey (Appendix 1) and a retrospective chart review (Appendix 5) to assess the rate of loss-to-follow-up of congenital heart patients after transition to adulthood and identify correlates of lost-to-follow-up.

3.2 Population and Sample:

All patients with structural heart disease who were transitioned from their pediatric cardiologist at the University of Saskatchewan to an adult cardiologist within a seven year period (2007-2014) were contacted for possible inclusion. They must have had at least one visit to pediatric cardiology between the ages of 13 and 17, indicating their heart disease was not cured in childhood.

Similar to previous studies assessing ACHD transition rates, we excluded patients who have died, those with Wolff-Parkinson-White syndrome and morphologically normal hearts, isolated cardiomyopathy, or pulmonary hypertension without structural heart disease. We also excluded patients who received a heart transplant in childhood as their follow-up care needs are more complex than the average ACHD patient.

Patients were identified as meeting the inclusion criteria from the pediatric cardiology database at the University of Saskatchewan, Royal University Hospital. This is the only facility in the province where pediatric cardiologists practice.

A pediatric cardiologist (Dr. Kakadekar or his nurse clinician, Marie Penner, who has been a part of the circle of care for these patients for many years) and/or Michelle Keir (MSc student) then contacted the potential participants using the last contact information on the pediatric chart. As this was often outdated, if we ended up speaking to the participant's parents, we informed them of the study and asked them either to have their child contact the study team, or provide contact information.

Telephone consent was requested to access the chart information and allowed us to call the participants and administer the survey. Participants could say yes to the chart review but opt out of the survey if they wished.

3.3 Data Collection Instruments and Variables

Once consent was obtained, the patient's charts were accessed in their various locations and the patient was called for the survey as appropriate.

A chart review was conducted from their pediatric charts (see Appendix 5). Patients who consented were contacted for a telephone survey (Appendix 1). The survey was tested by first inviting adult congenital heart patients, transitioned prior

to 1999, and content experts not involved in the study team to participate. They were asked to provide feedback on the format and questions, and it was modified accordingly. If it was determined, through the course of the study, that a patient was not receiving adequate follow-up, the general cardiologists in Saskatoon agreed to take them on as patients and ensure they receive adequate care if a gap was identified.

3.4 Data Analysis Procedures:

Data was analyzed using SPSS. To determine sociodemographic, clinical, and patient factors associated with a lack of appropriate follow-up, multiple logistic regression was performed using a backward stepwise method. Results were reported as odds ratios with 95 per cent confidence intervals. A p-value of 0.05 was used as a cut-off for statistical significance.

3.5 Knowledge Translation

Findings of this research will be shared with all pediatric and adult cardiologists in the province of Saskatchewan. This could potentially lead to improvements in program planning for this unique patient group. A scientific manuscript will be written and submitted for publication in a peer-reviewed journal. An abstract will also be submitted to ACHD related meetings to present the findings to the broader scientific community.

3.6 Anticipated Ethical Issues

The first ethical issue is the contact of patients for consent to participate in the study. The protocol was approved by the Research Ethics Board at the University of Saskatchewan and it was agreed that the marginal harm to patients of having their data accessed is outweighed by the potential benefit of the study.

The second ethical issue that could have arisen is the issue of loss-to-follow-up. It was plausible that, during the course of the research, we may identify patients who were lost to care. If these patients were identified, they were encouraged to contact their previous cardiologist and notify them of the care gap as well as book an appointment for follow-up.

3.7 Significance of the Study

As more patients with congenital heart disease reach adulthood, their care needs become paramount. Several studies have been undertaken to assess these needs in large tertiary care centres with trained ACHD professionals. Little research has been done in centres like Saskatchewan with non-existent frameworks for transition and lifelong care. This research will provide the basis for improved program planning in the province of Saskatchewan for children with congenital heart disease.

4.0 Results

A review of the pediatric cardiology database at the University of Saskatchewan identified 106 young adults who should have transferred to adult care between 2007 and 2014 (Figure 4.1). Each of these young adults had cardiac conditions requiring lifelong cardiac follow-up. Telephone contact with all 106 potential participants was attempted. Thirteen potential participants were reached by telephone but declined to complete the survey. Eleven patients were unable to complete the telephone survey; two were deceased, one was unwell and admitted to hospital, seven were unable to complete the survey according to their parents as they had significant learning disabilities or were non-verbal, and one patient had a hearing impairment which precluded the performance of a telephone survey. Twenty-seven possible participants were unreachable as the contact phone number on their pediatric file was out of service. In the case of 23 potential participants, telephone messages were left with either the patient or their parents regarding the survey but messages were never returned. Ultimately, 32 patients (30% response rate) consented to participate and completed the telephone survey.

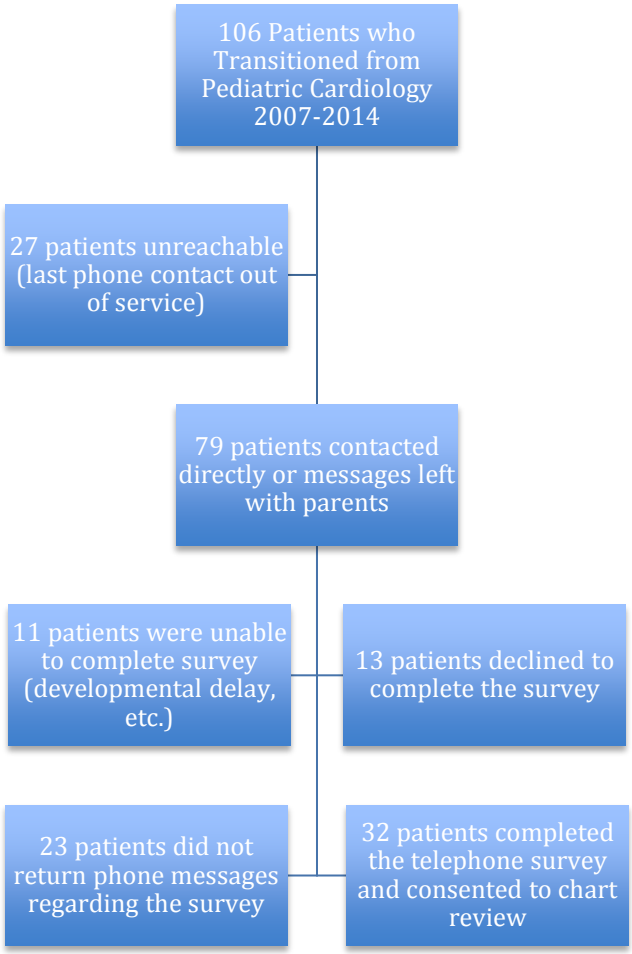


Figure 4.1 Potential participants and survey response rate

4.1 Demographics of Respondents

The majority of respondents to the telephone survey were male (n=19, 59%). All of the participants were born between 1990 and 1997 and the average age of the respondents at the time of the survey was 22 +/- 2 years. All but one of the participants, who immigrated to Canada from the Phillipines at age 14, were born in Canada and their first language was English. None of the participants reported asking for an interpreter during medical appointments as they were all comfortable

communicating with health care practitioners in English. Most of the respondents self-identified as Caucasian (n=26, 81%) while three identified as Aboriginal (9%), two identified as South Asian (6%), and one identified as Black (3%).

4.2 Education Level of Respondents

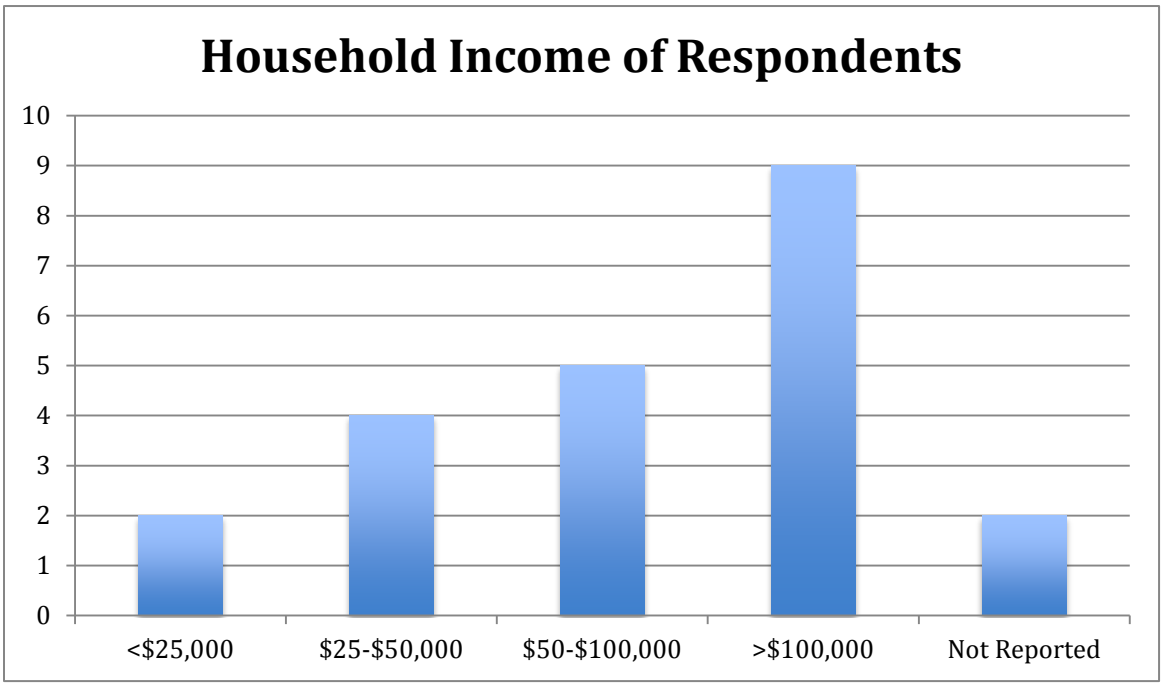
The majority of respondents (n= 27, 84%) reported having no disabilities (learning, physical, visual, or hearing). Four identified as having learning disabilities and only one reported a physical disability (residual arm weakness due to a pediatric stroke). All respondents had completed high school with the exception of one who was finishing his last year at the time of the survey. Two respondents disclosed that they graduated from a modified educational program due to their learning disabilities. Ten respondents (31%) had completed university or college degrees and one reported achieving an advanced degree. Six (19%) were currently attending college or university in pursuit of a degree.

4.3 Socioeconomic Status of Respondents

Only four respondents (13%) were neither employed nor full-time students. Approximately one-third (n=11, 34%) was working full-time for pay. The remainder was either working part-time for pay (n=12, 38%) or identified as full-time students (n= 5, 16%). Many of the young adults surveyed performed manual labour (heavy equipment operation, waste disposal, etc.). Several (n=4, 13%) worked in family businesses which they identified as providing flexibility and medical leave as required. Very few participants (n=4, 13%) reported ever being unemployed because of their congenital heart disease with three saying they could not sustain employment due to medical appointments and surgeries. One respondent cited his learning disability and overall health status as the reasons for his unemployment.

As a marker of socioeconomic status, we queried the respondents regarding food insecurity. In response to the question "How often in the past year was the following statement true? The food that you and other household members bought just didn't last and there wasn't any money to buy more," only three respondents (9%) signaled food insecurity by answering that the statement was "sometimes true". Many of the young adults surveyed reported living in a home owned by their parents (n=17, 53%). Three respondents lived in homes they had purchased and the remainder (n=12, 38%) was renting. The median reported income was between $50,000 to $100,000 Canadian dollars per year and that supported an average of 3.0 +/- 1.7 household members (Figure 4.2).

Figure 4.2 Median Reported Household Income of Participants with ACHD



As a measure of travel distance from home to a tertiary cardiac centre, we collected postal codes from the pediatric charts of all respondents. About half of the respondents lived in major cities in Saskatchewan (n=17, 53%) while the rest grew up in rural communities (n=15, 47%). The mean distance from pediatric address to the adult cardiologist to whom the patient was transitioned was 62.1 +/- 87.6 kilometres. The majority of respondents were raised in two parent households (n=27, 84%) while the remainder (n=5, 16%) was raised in single parent households. Only one participant was raised by adopted parents while the rest were raised by biological parent(s).

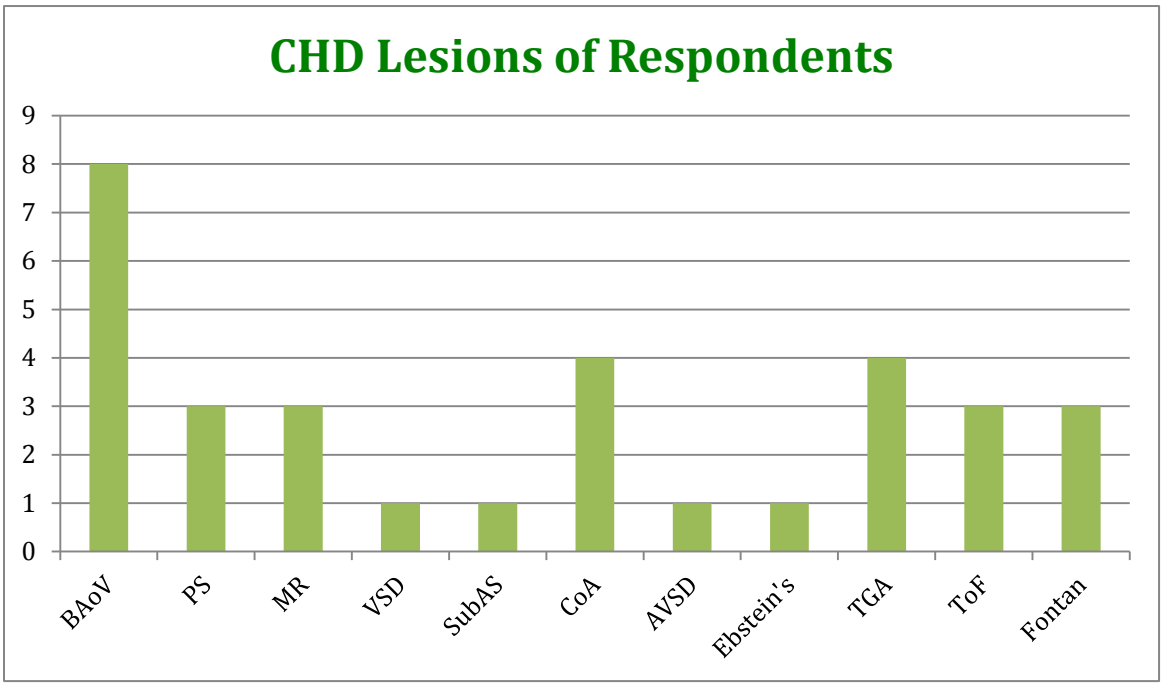
4.4 Substance Use

The majority of young adults surveyed (n=28, 88%) were lifelong non-smokers. One was an ex-smoker and three were actively smoking. Twenty-five of respondents (78%) drank alcohol although none reported exceeding the Canadian low-risk drinking guidelines[70]. One-quarter of respondents (n=8, 25%) disclosed the use of illicit drugs (drugs not prescribed by a physician). The majority of those who used street drugs had only used marijuana (n=6, 75%) but two respondents had experimented with other drugs including ecstasy, acid, mushrooms, and cocaine. Only one respondent was a daily marijuana user.

4.5 Congenital Heart Disease Anatomy

The distribution of anatomical lesions present in survey respondents is presented in Figure 4.3. Sixty-nine per cent of participants (n=22) had CHD of moderate-severe complexity while the remainder (n=10, 31%) had simple CHD.

Figure 4.3 CHD Lesions of Respondents



BAoV = bicuspid aortic valve, PS = pulmonary stenosis, MR = mitral regurgitation, VSD = ventricular septal defect, SubAS = subaortic stenosis, CoA = coarctation of the aorta, AVSD = atrioventricular septal defect, Ebstein's = Ebstein's anomaly of the tricuspid valve, TGA = transposition of the great arteries, ToF = tetralogy of Fallot, Fontan = Fontan circulation/operation

The mean number of open cardiosurgical procedures was 1.6 +/- 1.3 per patient. More than half of the respondents (n=18, 56%) were taking at least one medication daily for their heart disease. Half of respondents (n=16) had congenital heart lesions that required antibiotics prior to dental procedures (endocarditis prophylaxis) as per 2014 American College of Cardiology Valvular Heart Disease Guidelines.[71]

4.6 Knowledge of Congenital Heart Disease

Twenty-five of the respondents (78%) knew the medical name of their original cardiac lesion. Although many of them knew the medical name of their congenital cardiac anatomy, less (n=20, 63%) were able to describe the anatomy in layman's terms. Even fewer (n=15, 47%) were able to provide the medical name or describe the cardiac surgeries they had undergone. Of the patients who were taking at least one cardiac medication, only slightly more than half (n=11, 61%) knew why they were taking the medication or its physiologic purpose. Only 72% (n=23) of patients were appropriately adhering to endocarditis prophylaxis guidelines based on their anatomy (taking antibiotics prior to dental procedures if recommended) (Table 4.1). Most of those who were not adhering to guidelines were continuing to take prophylactic antibiotics despite no longer requiring them (n=7, 78%). Of more concern, two patients with artificial heart valves, who should be taking antibiotics prior to dental procedures, were not aware that it was still required and were not adhering to the practice.

Table 4.1 Appropriate use of antibiotic prophylaxis prior to dental procedures

	Takes Endocarditis Prophylaxis	**Does Not Take Endocarditis Prophylaxis**
Should Take Endocarditis Prophylaxis	14	2
Should Not Take Endocarditis Prophylaxis	7	9

4.7 *Reproductive Health*

None of the respondents had children, although one of the female respondents was in her first trimester of her first pregnancy at the time of the survey. Of the thirteen women surveyed, three had a history of pregnancy (23%). The two women who were not currently pregnant had both undergone terminations; one for personal reasons and the other because she developed heart failure in her second trimester and therapeutic termination was recommended by her cardiologist. Of the women who had been pregnant, all three reported having a discussion with their cardiologist prior to pregnancy delineating the cardiac risks. Of the women who had not been pregnant (n=10), only 50% reported that their cardiologist had initiated a conversation about possible future pregnancy and the risks. Of the 12 women not pregnant at the time of the survey, half of them were using some form of birth control (n=6, 50%). Three were taking oral contraceptives, one had an intrauterine device (IUD) in place, one was using barrier methods (condoms), and one had undergone a tubal ligation.

4.8 Health Care Utilization

Almost all of the respondents, with the exception of two, had regular follow-up with their family physician (general practitioner). All of the respondents, with the exception of one, were under the care of a cardiologist. Only twelve patients (38%) were under the care of a cardiologist with specialized training in ACHD. Of

the twenty-two patients with moderate to severe complexity of their CHD, less than one-third (n=7, 32%) were appropriately under the care of an ACHD specialized cardiologist. The remainder (n=15, 68%) were being cared for by general cardiologists with no specialized training in ACHD. Overall, only 69% (n=22) of respondents were receiving appropriate guideline-based follow-up for their ACHD.[3]

On self-report measures of successful transition and cardiology follow-up, very few participants reported attending clinic appointments without a parent or guardian as a teenager (n=4, 13%). Six patients (19%) reported missing clinic appointments with their cardiologist. Reasons cited for missing scheduled cardiology appointments included unstable housing leading to missed appointment letters, difficulty scheduling clinic visits with busy cardiology practitioners, and poor weather making driving long distances to clinic appointments difficult. One female patient cited significant anxiety about long-term cardiac health as a reason for many missed or rescheduled cardiology appointments.

Overall, three patients (9%) were lost-to-follow-up from cardiology for more than two years at the time of the survey. All three lost patients were male. One patient had Williams syndrome and developmental delay and was unsure why he had not seen his cardiologist in some time. The other two reported that they had not been contacted by their cardiologist for an appointment and had not followed up. One of the patients with Ebstein's anomaly characterized his understanding as such:

"I thought they would contact me and, since they didn't, I figured I was fine and dandy … good to go!"

All three respondents who reported being lost to follow-up were informed that they did, indeed, need lifelong cardiology follow-up and were encouraged to contact their primary cardiologists to schedule an appointment.

4.9 Social Supports and Self-Care

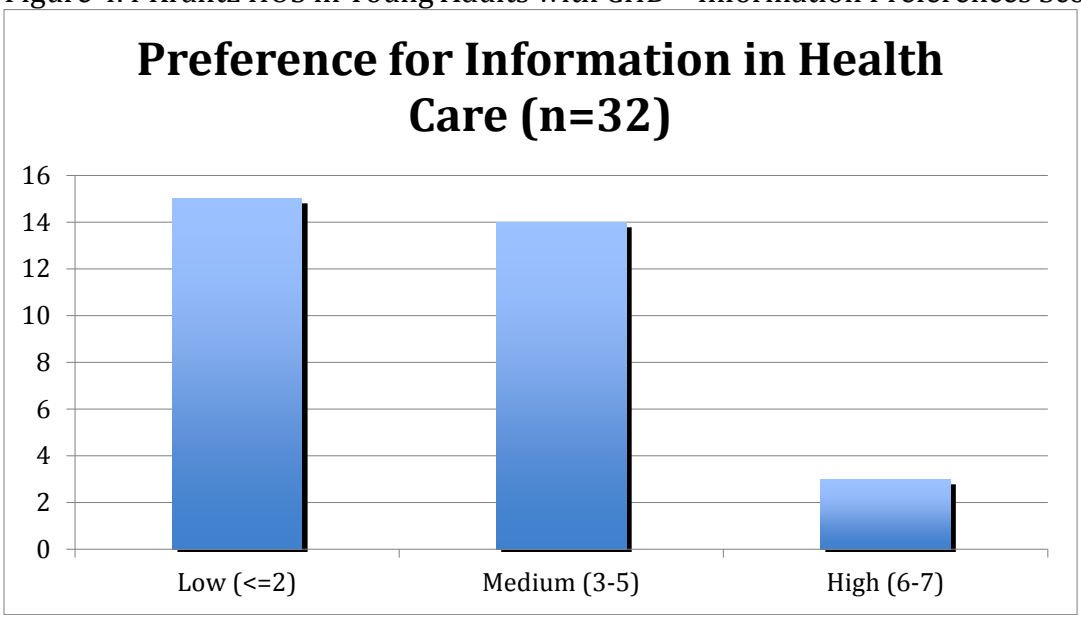
Few of the respondents were married or had a common-law spouse at the time of the survey (n=4, 13%). We performed the ENRICHD Social Support Inventory Survey to assess perceived social supports among young adults with ACHD.[72] Only one patient's survey results indicated that she had low perceived social support, while the remainder reported adequate social support. We also performed the PHQ-9 depression screen and only two patients met criteria for major depressive disorder (MDD).[73]

4.10 Krantz Health Opinion Survey

All 32 participants agreed to complete the Krantz HOS survey. Seventy-five per cent of participants (n=24) received a low overall score on the Krantz HOS survey (6 or less). The remainder (n=8, 25%) received a medium score. None of the patients surveyed received a high score suggestive of health care autonomy. When stratified based on preferences for receipt of medical information and self-care behaviours (Figures 4.4 and 4.5), more participants (n=17, 53%) indicated a

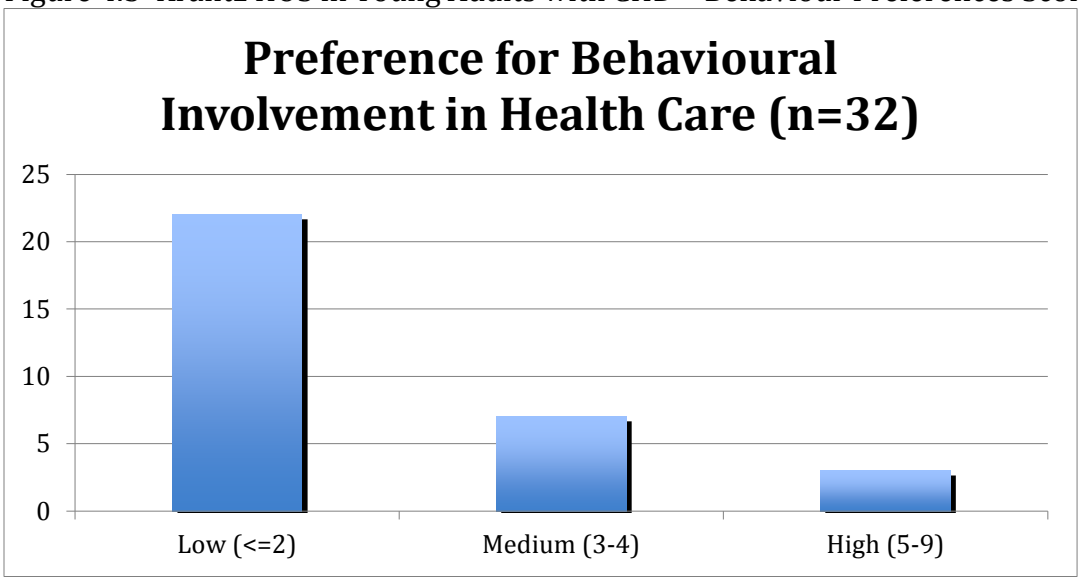
preference for receiving information about their health than those who wanted to be actively involved in decision-making about their health (n=10, 31%).

Figure 4.4 Krantz HOS in Young Adults with CHD – Information Preferences Score



Note: A low score on the Krantz HOS Information score (2 or less) indicates a preference to not receive detailed information about health and a predilection to not ask questions of health care practitioners

Figure 4.5 Krantz HOS in Young Adults with CHD – Behaviour Preferences Score



Note: A low score on the Krantz HOS Behaviour score (2 or less) indicates a preference to leave health care decisions to medical practitioners and not participate in care decisions.

4.11 Qualitative Assessment of Transition Process

At the conclusion of the telephone survey, participants were invited to provide comments they thought would be useful for improving the transition process for teenagers with CHD. They were asked the open-ended question; "Do you have any comments or suggestions to improve the transition process?"

Many respondents (n=22, 69%) felt that the transition process was smooth and could not think of any measures that may have improved their experience. One participant noted that the transition process was tougher for his parents than it was for him and expressed relief that he could now take responsibility for his own health decisions. He felt that an education session for parents around the time of transition explaining the importance of health care autonomy for their young adult child

would have been useful. Another young woman expressed pride in the fact that she now managed her own heart disease and considered herself an advocate for her own care.

Other respondents noted that the time between their last pediatric appointment and their first adult appointment was a time of uncertainty. One young woman wished she had been clearly been provided with the contact information of her new adult cardiologist as she was unsure what to do when she experienced cardiac symptoms during the transition period. Another young man expressed frustration that an intervention (open heart surgery for valve replacement) was planned during the time of transition. His surgery was planned and arranged by the pediatric team and his post-surgical follow-up was completed by his new adult cardiologist. He suggested that all planned interventions occur prior to transition or be postponed, if possible, to adulthood.

Seven respondents (22%) noted that they feel rushed during appointments with their adult cardiologist and that pediatric providers had more time for detailed explanation and relationship building. One young woman noted that she feels out of place in her adult cardiologists' office because "everyone else is 80 plus (years old) in the clinic". She wished that she could see a dedicated ACHD cardiologist as she would like to be in a clinic with "a younger clientele". Another young woman said she feels like "just a number in the system" in adult care as opposed to pediatric cardiology where she had a more congenial relationship with her providers. One man felt that his adult cardiologist was too busy and that his administrative staff was overwhelmed. He related an episode where his adult cardiologists' office failed

to schedule a planned follow-up appointment; “we had to phone and follow-up. I would have got lost if it wasn’t for my Mom.” Another young man attributed his ongoing follow-up to his own persistence; “I feel like I’ve gotten lost”.

One respondent expressed frustration with the health care system overall. He felt that other health care providers should be more educated regarding congenital heart disease and its possible sequelae. He related an episode as a teenager when he presented to a walk-in clinic with fevers, later determined to be a result of infective endocarditis, and was told he had contracted a common cold. He had no criticisms of his pediatric or adult cardiology teams but felt the walk-in practitioner should have recognized his serious condition and that the delay in adequate treatment had negatively affected his overall health.

4. 12 Statistical Analysis

A logisitic regression model was attempted to determine variables that were associated with the outcome of interest. Data were analyzed using IBM SPSS Statistics, versions 24 and 25. The outcome of interest was a dichotomous variable defined as loss to cardiology follow-up for more than two years and/or missed clinical appointments. A bivariate analysis was performed to assess for possible correlation between risk factors and the outcome of interest. Potential risk factors assessed were; biological sex, distance of transition address to cardiology clinic, age at survey, racial identity, the presence of disability (visual, learning, or physical), level of education, family income, lesion complexity, number of surgeries, the

utilization of medications, low perceived social supports, low Krantz HOS score, and the presence of a major depressive disorder. Population characteristics stratified by primary outcome are presented in Table 4.2.

Table 4.2 Population characteristics of ACHD patient surveyed stratified by primary outcome

	Participants lost to follow-up>2 years and/or missed cardiology appointments (Cases, n=8)	Participants with continuous cardiology follow-up (Controls, n=24)
Age (Mean +/- SE)	23.0 +/- 0.8	21.5 +/- 0.4
Male Sex (n (%))	4 (50%)	15 (62.5%)
Transition Distance in kilometres (Mean +/- SE)	38.6 +/- 25.2	70.0 +/- 18.9
Self-Identified Race/Ethnicity (n, %)	Caucasian (6, 75%) Aboriginal (2, 25%) Other (0, 0%)	Caucasian (20, 83.3%) Aboriginal (1, 4.2%) Other (3, 12.5%)
Presence of Disability (n (%))	1 (12.5%)	5 (20.8%)
Highest Education Level Achieved (n, %)	Advanced degree (1, 12.5%) College/University degree (3, 37.5%) Some College/University (1, 12.5%) Completed High School (3, 37.5%) Less than High School (0,0%)	Advanced degree (0, 0%) College/University degree (7, 29.2%) Some College/University (9, 37.5%) Completed High School (7, 29.2%) Less than High School (1, 4.2%)
Family Income in CAD$ (n, %)	>100,000 (1, 12.5%) 50-100,000 (3, 37.5%) 25 – 50,000 (1, 12.5%) <25,000 (1, 12.5%)	>100,000 (7, 29.2%) 50-100,000 (12, 50%) 25 – 50,000 (3, 12.5%) <25,000 (1, 4.2%)
ACHD Lesion Complexity (n, %)	Simple (2, 25%) Moderate (4, 50%) Great (2, 25%) Great+ (0, 0%)	Simple (8, 33.3%) Moderate (9, 37.5%) Great (4, 16.7%) Great+ (3, 12.5%)
Number of Cardiac Surgeries (Mean +/- SE)	1.75 +/- 0.7	1.5 +/- 0.2
Taking Cardiac Medications (n (%))	3 (37.5%)	15 (62.5%)
Low Perceived Social Support (n, %)	1 (12.5%)	0 (0%)
Krantz Health Opinion Survey Score	Low (5, 62.5%) Medium (3, 37.5%) High (0, 0%)	Low (21, 87.5%) Medium (3, 12.5%) High (0, 0%)

Meets Criteria for Major Depressive Disorder (n (%))	1 (12.5%)	1 (4.2%)
Cared for by an ACHD trained cardiologist (n(%))	2 (25%)	10 (41.7%)

4.12.2 Bivariate analysis results for each of the 14 potential risk factors predicting the primary outcome of interest (lost to cardiology follow-up for >2 years and/or missed cardiology appointments) are provided below in Table 4.3.

The only variables that were potentially predictive of the primary outcome were age at survey, self-identification as Aboriginal, taking cardiac medications, and medium Krantz health opinion score. Since there were no patients with low perceived social support that were lost to cardiology follow-up, the odds ratio for this variable was indeterminate (denominator of 2x2 table was zero). A multivariate analysis was attempted but, due to the low sample size and the multiple number of comparisons required, the results were considered invalid and will not be reported. Therefore, we can only suggest that variables identified via bivariate analysis may have some correlation with the primary outcome of interest but no definitive conclusions can be made based on the data collected due to the small sample size.

Table 4.3 Bivariate analysis results for each of the 14 potential risk factors.

Variable	Odds Ratio	95% CI of OR	P-value
Age	1.440	(0.931, 2.228)	0.101
Sex	0.600	(0.120, 3.012)	0.533
Race Aboriginal Other	 6.667 0.0	 (0.511, 86.929) (0, …)	0.351 0.148 0.999
Transition Distance	0.995	(0.984, 1.006)	0.385
Education Level (no college/university)	0.833	(0.1578, 4.4007)	0.8300
Family Income < $25,000 CAD	3.500	(0.145, 84.694)	0.441
Simple Lesion Complexity	0.375	(0.035, 3.999)	0.417
Disability	0.543	(0.054, 5.498)	0.601
Number of Cardiac Surgeries	1.129	(0.617, 2.064)	0.695
Taking Cardiac Medications	0.360	(0.069, 1.880)	0.217
Low Perceived Social Support	Indeterminate	Indeterminate	Indeterminate
Krantz Health Opinion Score (Medium with Low as reference category)	4.200	(0.645, 27.362)	0.133
Major Depressive Disorder	3.286	(0.181, 59.599)	0.399
Cared for by an ACHD Cardiologist	2.143	(0.356, 12.890)	0.399

5.0 Discussion

5.1 *Major Findings*

This study reveals several novel findings regarding the care and follow-up of young adults with CHD.

Our primary outcome measure was success of transition measured by self-reported loss to follow-up and/or missed appointments with adult cardiologists. The only statistically significant predictors of the primary outcome were older age and no need for cardiac medications. This finding is interesting in that it suggests we may be overestimating the transition period and failing to recognize ongoing attrition as ACHD patients age. Although many patients are lost during the transition period, frustrations with the health care system and poor administrative supports may cause patients to become lost at an older age. This finding, in addition to qualitative comments from participants that they find adult care paradigms less hospitable, suggests we should be improving communication regarding the importance of long-term follow-up in the adult phase of care. We also found that a lack of cardiac medications, indicating no pressing need to meet with adult cardiologists to receive prescription renewals, also had a statistically significant correlation with loss to follow-up. This also indicates an ongoing attrition due to dissatisfaction with the adult paradigm of care.

While travel distance to adult care was not statistically significantly correlated with failed transition, several patients verbally cited weather and travel as reasons for multiple missed appointments. This suggests that distance to care providers is an ongoing burden on patients in rural areas and requires further

study. We found that 47% of patients surveyed lived in rural communities with no tertiary care centres and that they travelled an average of 62km to see their cardiologist. Although distance to cardiology care was not statistically significantly associated with loss-to-follow-up or missed appointments, our sample size was likely insufficient to detect a significant difference. Previous research assessing distance to care and transition success has been performed in small European countries or large metropoleis.[14,16] It is clear that travelling long distances to access care is a financial barrier. This study highlights the need for future research assessing cardiac outcomes in ACHD patients who live in remote rural areas.

When assessing social stressors, we identified that 13% of patients surveyed reported either currently being unemployed because of their congenital heart disease or having been unemployed in the past as a result of their heart. An additional interesting finding was that 13% of patients were employed in family businesses and stated that the reason was flexibility and understanding regarding their underlying congenital heart defect and limitations. Despite a high level of educational attainment in our cohort, and an overall high level of employment compared to previously reported cohorts[38], this finding highlights the difficulty of living with congenital heart disease which is perhaps not captured by quality of life scoring alone.[41] Previously published qualitative research has demonstrated that congenital heart disease affects all aspects of the life course of patients, including decisions regarding relationships and career.[74] This highlights the importance of care practitioners understanding the social burden of chronic disease on young patients. Frequent doctor's appointments and tests, often requiring travel and time

off work or school, are much more disruptive in the lives of young adults than they are in the lives of the average cardiology patient who is in their sixth or seventh decade of life. Perhaps the traditional care paradigms of health care should be modified to accommodate the needs of these young patients. Suggestions include flexible appointment times (evenings and weekends) and telemedicine as a means to minimize travel.

In addition, this is the first study of congenital heart patients to query food insecurity. We found that three respondents, or nine per cent, signaled food insecurity when asked how often they were unable to afford needed food in the year prior. ACHD patients are known to have lower incomes than age-matched peers but markers of poverty have never truly been assessed.[12] When assessing barriers to transition and long-term follow-up, care practitioners often naively assume that cardiovascular health and longevity should be the number one priority for our patients. When viewed through the lens of socioeconomic barriers, if a patient is unable to purchase food because their financial situation is so dire, it is easy to see how long-term health and preventive medicine can be a low priority for them. How can we expect patients to prioritize travel to medical appointments over basic sustenance? This finding highlights the importance of screening for food insecurity in general clinical practice. Although it is not traditionally the role of the physician to assess for and/or alleviate poverty, evidence reveals that food insecurity is linked to higher health care costs.[75] Perhaps referring patients to assistance programs and providing resources may be of more long-term benefit to some patients than

traditional health counselling, education about their congenital heart disease, and cardiac investigations.[75]

We also found that one-quarter of the young adults with ACHD we surveyed (25%, n=8) self-reported use of illicit substances. This is in line with previous Canadian research[44], but requires more detailed investigation. The majority of respondents to our survey who used illicit drugs had only used marijuana. The consequence of the legalization of marijuana in Canada and its long-term health effects on patients with ACHD has yet to be studied.

Another interesting component of our study was our assessment of CHD knowledge. Similar to previous studies, we found that overall knowledge regarding CHD was poor among those surveyed; only 61% of those taking cardiac medications could state the reason they were taking them and several could not name their congenital heart defect.[54] In addition, adherence to endocarditis prophylaxis was poor. Many patients were still taking antibiotics prior to dental procedures despite no longer requiring them and two patients were not taking antibiotics when they, in fact, required them for endocarditis prevention. Clearly, when guidelines change over time, it sows confusion among patients. This highlights the need for long-term consistency in our approach to management and clear communication with patients when management paradigms change.

Perhaps the most prescient findings of this study were the results of the Krantz HOS survey. An assessment of knowledge preferences in ACHD patients has not previously been reported in the literature. We found that only half of ACHD patients surveyed wanted to receive information about their health and less than a

third wanted to be involved in decision-making. This is both surprising and extremely important information when assessing ways to improve follow-up in ACHD. There is ongoing research trying to educate patients about their ACHD and assessing whether improved knowledge improves transition. The results of our survey indicate that this research is likely futile. We know that patients with ACHD have significant anxiety, and even medical post-traumatic stress disorder[50]. Trying to force education regarding anatomy and medical terminology on patients with medical PTSD may be counter-productive and drive them further away from care. Instead, partnering with patients by highlighting symptoms they can watch for, discussing the reasons for long-term follow-up, and making them feel safe and cared for may have better effect in the long-term.

The results of the Krantz HOS survey raise another important question. Has the pediatric paradigm of cardiac care institutionalized young adults and prevented them from achieving health care autonomy? We know that the majority of adult patients have fond memories of and implicit trust in their pediatric cardiologists and nurse clinicians.[74] Does heavy contact with the medical system during the formative years create an external locus of control in ACHD patients and convince them that they should defer all medical decisions to professionals? How can this cycle be broken? Perhaps, instead of focusing on the medical education of our patients, aiming to teach health care autonomy would lead to better transition success. Further qualitative and quantitative research in this area is warranted.

Another major finding of this study was the lack of guideline-based care provided to patients with ACHD in Saskatchewan. Recommendations suggest that all

patients with CHD of moderate to severe complexity be followed by a cardiologist with specialized training in ACHD (usually an additional one or two year fellowship in a centre with a large volume of ACHD patients)[3]. There is only one cardiologist in Saskatchewan with the requisite training who practices in Regina. Not surprisingly, therefore, only 32% of patients surveyed with moderate-complex CHD were appropriately followed by an ACHD cardiologist. The remainder was followed by general cardiologists. This may have serious ramifications on the care and long-term outcomes of ACHD patients. There is clear evidence that specialized ACHD care improves outcomes and mortality.[20] One could then argue that patients with ACHD in Saskatchewan are at higher risk of mortality because of a lack of appropriately trained care providers. A lack of appropriately trained adult cardiologists to whom patients can be transferred is an identified barrier to transition as well. Several survey respondents reported feeling uncomfortable in their adult cardiologists' offices because they were not within their primary practice demographic. Therefore, this study provides evidence that appropriately trained ACHD cardiologists are needed in Saskatchewan to facilitate transition and also to improve mortality in adults with CHD.

Additionally, we identified that only 50% of women with ACHD were counseled regarding the cardiac risks of pregnancy to themselves and the risk of congenital heart disease in offspring. This is another symptom of a lack of cardiologists appropriately trained in cardiac disorders of pregnancy in the province of Saskatchewan. There is little emphasis on cardiac disorders of pregnancy during general cardiology training but most ACHD fellowship programs

have obstetrical medicine rotations and focus on these problems. ACHD cardiologists, as we deal with young patients, often manage cardiac complications of pregnancy and are comfortable counseling young women about risks. The lack of appropriate information provided to women with ACHD in Saskatchewan raises an equity issue as it suggests that women with ACHD receive sub-standard care surrounding pregnancy due to a dearth of appropriately educated providers.

Finally, we also found that a major reason for loss to follow-up and missed appointments cited by patients was administrative. Two patients reported difficulty scheduling clinic visits due to busy cardiologists and another one reported that he was never contacted for a follow-up appointment so he assumed he did not require follow-up. This has been cited previously and suggests the "out of sight, out of mind" attitude of CHD patients.[13] It is also a symptom of the lack of health care autonomy we identified with the Krantz HOS survey. If we know that patients with ACHD will likely not take matters into their own hands and ensure a return to care, we can then advocate for better tracking systems and databases to ensure that the health care system does not lose them. This qualitative finding suggests that we are failing patients with our current care paradigm and that we need novel ways of communication and tracking to keep them in care.

5.2 *Generalizability*

An important component to the generalizability of our findings is a comparison to both the general Canadian youth population and to previously

reported cohorts of young adults with ACHD. Unfortunately, since we had little clinical or demographic information regarding non-respondents to our survey, a major limitation of the study is the inability to compare non-respondents to respondents.

For unclear reasons, we had a larger proportion of male respondents than female (59%). This is different than the average biological sex distribution of ACHD with the prevalence of CHD being higher in female adults than male adults.[2] In addition, 81% of our respondents were Caucasian which is a higher percentage than the average population of Saskatchewan.[76] According to the latest 2016 Canadian census, 16.6% of the Saskatchewan population identifies as Aboriginal and 11.8% is of African, Latin American, or Asian origins.[76] Also, all of our participants except one spoke English as their first language. Therefore, our results are not necessarily generalizable to the entire Saskatchewan population. This also highlights the difficulties patients without proficiency in English face in accessing the health care system in their non-native language.

Additionally, 50% of our respondents had either completed, or were in the process, of completing a University degree. This suggests that participants had a higher education level than the average population of Saskatchewan and our results may not be generalizable to those with lower educational attainment.[76] This is also in line with the presumption that people with higher educational attainment are more likely to participate in invited research.

Interestingly, respondents to our survey were less likely to use cannabis than the general Canadian population (25% vs. 41%) and less likely to use other illicit

drugs (6% vs. 11%).[77] This may result from a higher level of general health literacy and consciousness among young adults with ACHD or from pediatric counseling to avoid substance abuse from allied health care professionals.

Although our participants were more likely to be Caucasian, better educated, and less likely to use substances than the general population of Saskatchewan, our cohort is similar to previously reported ACHD cohorts from Canada. The largest described dataset of ACHD patients in Canada comes from administrative data in Quebec.[2] Our cohort had similar numbers of patients with complex anatomical lesions as that in the general Canadian ACHD population.[2] As stated previously, we had more men in our cohort than in the average ACHD population (59% vs. 43%).[2] There is no previous Canadian data regarding educational level and socioeconomic status of adults with ACHD. Despite the lack of Canadian data, the proportion of our cohort that had attended college or university is similar to that reported in a Belgian cohort of ACHD patients (50% vs. 47%).[16] We also found similar rates of substance abuse to a previously reported cohort of young adults in Ontario with ACHD.[14] Therefore, although the average participant in our cohort is different from the average Saskatchewan young adult, he/she would be quite similar to those in previously-reported cohorts of young adults with ACHD. We can, therefore, postulate that our results generalize to the average young adult with ACHD.

5.3 Limitations

Our study highlights the difficulty in tracking patients as they transition from pediatric to adult care paradigms. A major limitation of this study was our participation rate as we were unable to contact many young adults who had transitioned from pediatric cardiology in Saskatchewan. Although the pediatric cardiology program in Saskatoon has an internal electronic database, once patients are transitioned to adult care, their electronic health information does not travel with them. Often, each hospital or clinic has its own internal database that does not easily communicate with others, making movement of patient information between facilities and health regions almost impossible. In addition, international classification of diseases (ICD) codes for congenital heart disease are notoriously vague and non-descript, making government and administrative databases used for billing and tracking health outcomes unreliable in this population[78].

Also, the transition from pediatric to adult care occurs at a period of general transition in the adolescent life cycle. Patients often move for work or to pursue higher education. In the cases of several patients we were ultimately able to contact, we were provided with accurate contact information by their parents. This suggests that those children with less stable social circumstances (parents who move frequently) were more difficult to contact and our sample is, therefore, skewed towards young adults with stable home lives during childhood.

Another complicating factor is the changing landscape of communication. For more than one quarter of potential participants, we were unable to make phone

contact as their land lines were out of service. This highlights the difficulty of communicating with young patients by traditional methods (letter mail and/or telephone). Research has shown that more primary care physicians are communicating with patients via text messaging and email.[79] Despite the risks of encroachment on their personal lives and a failure to protect the confidentiality of health information, 70% of physicians in Switzerland reported communicating with their patients via non-traditional means.[79] Our study highlights the urgent need for health systems to address the current technology gap between modern communication and methods for privacy protection.

Another limitation of this study was the method of the survey. By choosing to perform a telephone survey, we acknowledge that patients with hearing disabilities and those with significant developmental disorders were necessarily excluded. The generalizability of our results to the population of ACHD that is not high functioning is, therefore, limited. The choice of survey methodology was made for feasibility as in person interviews would place a prohibitive burden on investigators and study participants.

5.4 Future Directions – Changing Practice and Further Research

By utilizing a comprehensive telephone survey to contact young adults with ACHD in Saskatchewan, we have identified several potential barriers to care in the province. Based on the previously published literature and the results of this study,

the following recommendations can be made to improve the care of young adults with ACHD in Saskatchewan;

1. The recruitment of appropriately trained ACHD cardiologists to Saskatchewan should be a priority in an effort to improve morbidity and mortality of patients with ACHD in the province.
2. A comprehensive database should be created to track ACHD patients as they transition from pediatric life to adult care. Liaison of this database with those of other provinces, since young adults frequently move, is paramount to ongoing follow-up and tracking of patients with ACHD.
3. A formalized transition program should be created which educates young patients on health care autonomy and the importance of lifelong cardiology follow-up. This could be facilitated by allied health care professionals and would include the creation of health passports.
4. Other allied health care practitioners in Saskatchewan (family physicians, emergency room physicians, nurses, social workers) should receive continuing education regarding the growing number of ACHD patients and their specific needs to help facilitate their adult care.

In addition to identifying gaps in care, this study has also identified areas for future research. As the majority of ACHD research occurs with patients living in large urban areas, research on the outcomes and care patterns of patients living in rural areas should be further studied prospectively. This line of inquiry is especially prescient in a geographically diverse nation like Canada.

Also, we have uniquely identified that patients with ACHD have poor health care autonomy and a preference to defer health decision making to care providers. Therefore, the utility of educating and encouraging ACHD patients to practice health care autonomy could be the focus of a prospective, randomized controlled trial with the primary outcomes being clinical adherence, ACHD morbidity, and mortality.

6.0 Conclusion

In summary, patients with ACHD in the province of Saskatchewan are living longer than ever before due to advances in pediatric surgical techniques and the efforts of pediatric cardiologists. Our sample size of patients who completed the survey was too small to statistically define clear barriers to care but our qualitative assessment of care, provided by the patients who participated in the survey, suggests that ACHD patients in Saskatchewan are not receiving guideline-based care. Young adult ACHD patients also exhibit a lack of health care autonomy. These factors combine to create a high risk of morbidity and mortality for young adults with ACHD in Saskatchewan.

7.0 References

1. Moons, P., Bovijn, L., Budts, W., Belmans, A., & Gewillig, M. (2010). Temporal trends in survival to adulthood among patients born with congenital heart disease from 1970 to 1992 in belgium.. *Circulation, 122*(22), 2231-2233.

2. Marelli, A. J., Mackie, A. S., Ionescu-Ittu, R., Rahme, E., & Pilote, L. (2007). Congenital heart disease in the general population: Changing prevalence and age distribution *Circulation, 115*, 163.

3. Warnes, C. A., Williams, R. G., Bashore, T. M., Child, J. S., Connolly, H. M., Dearani, J. A., et al. (2008). ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: A report of the american college of Cardiology/American heart association task force on practice guidelines (writing committee to develop guidelines on the management of adults with congenital heart disease): Developed in collaboration with the american society of echocardiography, heart rhythm society, international society for adult congenital heart disease, society for cardiovascular angiography and interventions, and society of thoracic surgeons *Circulation, 118*(23), 714-833.

4. Fernandes, S., Khairy, P., Fishman, L., Melvin, P., O'Sullivan-Oliveira, J., Sawicki, G., et al. (2012). Referral patterns and perceived barriers to adult congenital heart disease care: Results of a survey of U.S. pediatric cardiologists. *Journal of the American College of Cardiology, 60*(23), 2411-2418.

5. Foster E, Graham Jr TP, Driscoll DJ, et al. Task force 2: special health care needs of adults with congenital heart disease. J Am Coll Cardiol 2001 Apr;37(5):1176–83.

6. van der Linde, D., Konings, E.E.M., Slager, M.A., Witsenburg, M., Helbing, W.A., Takkenberg, J.J.M., Roos-Hesselink, J.W. (2011). Birth prevalence of congenital heart disease worldwide: A systematic review and meta-analysis. *Journal of the American College of Cardiology, 58*(21): 2241-2247.

7. Samanek, M. (1992). Children with congenital heart disease: Probability of natural survival. *Pediatric Cardiology, 13*(3): 152-158.

8. Higashi, H., Barendregt, J.J., Kassebaum, N.J., Weiser, T.G., Bickler, S.W., Vos, T. (2015). The burden of selected congenital anomalies amenable to surgery in low and middle-income regions: cleft lip and palate, congenital heart anomalies and neural tube defects. *Archives of Disease in Childhood, 100,* 233-238.

9. Khairy, P., Ionescu-Ittu, R., Mackie, A.S., Abrahamowicz, M., Pilote, L., Marelli, A.J. (2010). Changing mortality in congenital heart disease. *Journal of the American College of Cardiology, 56*(14): 1149-1157.

10. Marelli, A.J., Ionescu-Ittu, R., Mackie, A.S., Guo, L., Dendukuri, N., Kaouache, M.

(2014). Lifetime prevalence of congenital heart disease in the general population from 2000 to 2010. *Circulation, 130,* 749-756.

11. Mackie, A.S. Pilote, L., Ionescu-Ittu, R., Rahme, E., Marelli, A.J. (2007). Health care resource utilization in adults with congenital heart disease. *American Journal of Cardiology, 99*, 839-843.

12. Zomer, A.C., Vaartjes, I., van der Velde, E.T., de Jong, H.M.Y., Konings, T.C., Wagenaar, L.J., …, Mulder, B.J.M. (2013). Heart failure admissions in adults with congenital heart disease; risk factors and prognosis. *International Journal of Cardiology, 168*, 2487-2493.

13. Wray, J., Frigiola, A., Bull, C. (2013). Loss to specialist follow-up in congenital heart disease; out of sight, out of mind. *Heart, 99,* 485-490.

14. Reid, G., Irvine, M., McCrindle, B., Sananes, R., Ritvo, P., Siu, S., Webb, G.D. (2004). Prevalence and correlates of successful transfer from pediatric to adult health care among a cohort of young adults with complex congenital heart defects. *Pediatrics, 113*(3 Pt1), e197-205.

15. Harbison, A.L., Grady, S., Chi, K., Fernandes, S.M. (2016). Provision of transition education and referral patterns from pediatric cardiology to adult cardiac care. *Pediatric Cardiology, 37*(2), 232-238.

16. Goossens, E., Stephani, I., Hilderson, D., Gewillig, M., Budts, W., Van Deyk, K., Moons, P. (2011). Transfer of adolescents with congenital heart disease from pediatric cardiology to adult heath care. *Journal of the American College of Cardiology, 57*(23): 2368-2374.

17. Mackie, A., Ionescu-Ittu, R., Therrien, J., Pilote, L., Abrahamowicz, M., Marelli, A. (2009). Children and adults with congenital heart disease lost to follow-up: Who and when?.. *Circulation, 120*(4), 302-309.

18. Gurvitz, M.Z., Inkelas, M., Lee, M., Stout, K., Escarce, J., Chang, R. (2007). Changes in hospitalization patterns among patients with congenital heart disease during the transition from adolescence to adulthood. *Journal of the American College of Cardiology, 49*(8): 875-882.

19. Wray, J., Frigiola, A., Bull, C. (2013). Loss to specialist follow-up in congenital heart disease; out of sight, out of mind. *Heart, 99,* 485-490.

20. Mylotte, D., Pilote, L., Ionescu-Ittu, R., Abrahamowicz, M., Khairy, P., Therrien, J., …, Marelli, A. (2014). Specialized adult congenital heart disease care: The impact of policy on mortality. *Circulation, 129,* 1804-1812.

21. Gurvitz, M., Valente, A., Broberg, C., Cook, S., Stout, K., Kay, J., et al. (2013) Prevalence and predictors of gaps in care among adult congenital heart disease patients;

HEART-ACHD. *Journal of the American College of Cardiology, 61*(21), 2180-2184.

22. Kempny, A., Diller, G.H., Dimopoulos, K., Alonso-Gonzalez, R., Uebing, A., Li, W., …, Gatzoulis, M.A. (2016). Determinants of outpatient clinic attendance amongst adults withcongenital heart disease and outcome. *International Journal of Cardiology, 203,* 245-250.

23. Yeung, E., Kay, J., Roosevelt, G.E., Brandon, M., Yetman, A.T. (2008). Lapse of care as a predictor for morbidity in adults with congenital heart disease. *International Journal of Cardiology, 125*, 62-65.

24. McCabe, N., Dunbar, S.B., Butler, J., Higgins, M., Book, W., Reilly, C. (2015). Antecedents of self-care in adults with congenital heart defects. *International Journal of Cardiology, 201,* 610-615.

25. Verheugt, C.L., Uiterwaal, C., van der Velde, E.T., Meijboom, F.J., Pieper, P.G., Vliegen, H.W., …, Mulder, B.J.M. (2008). Gender and outcome in adult congenital heart disease. *Circulation, 118,* 26-32.

26. Wacker, A., Kaemmerer, H., Hollweck, R., Hauser, M., Deutsch, M.A., Brodherr-Heberlein, S., …, Hess, J. (2005). Outcome of operated and unoperated adults with congenital cardiac disease lost to follow-up for more than five years. *American Journal of Cardiology, 95*, 776-779.

27. Beauchesne, L, Therrien, J., Alvarez, N., Bergin, L, Burggraf, G., Chetaille, P., …, Marelli, A. (2012). Structure and process measures of quality of care in adult congenital heart disease patients: a pan-Canadian study. *International Journal of Cardiology,* 157, 70-74.

28. Fernandes, S., Khairy, P., Fishman, L., Melvin, P., O'Sullivan-Oliveira, J., Sawicki, G., …, Landzberg, MJ. (2012). Referral patterns and perceived barriers to adult congenital heart disease care: Results of a survey of U.S. pediatric cardiologists. *Journal of the American College of Cardiology, 60*(23), 2411-2418.

29. Nguyen, L.T., Maul, T.M., Hindes, M., Daniels, C.J., Gurvitz, M., Cook, S.C. (2015). Current and future status of adult congenital training in North America. *American Journal of Cardiology, 115*, 1151-1153.

30. Deanfield, J., Thaulow, E., Warnes, C., Webb, G., Kolbel, F., Hoffman, A., …, Trappe, H. Management of grown up congenital heart disease: The task force on the management of grown up congenital heart disease of the European Society of Cardiology. *European Heart Journal* (2003): 24, 1035-1084.

31. Vasconcelos Amaral, F.T., Manso, P.H., Schmidt, A., Sgarbieri, R.N., de Andrade Vincente, W.V., Carbone Junior, C., Somerville, J. (2015). Recommendations for starting a grown-up congenital heart disease (GUCH) unit. *Brazilian Journal of Cardiovascular Surgery, 30*(3); 373-379.

32. Silversides CK, Marelli A, Beauchesne L, et al. Canadian Cardiovascular Society 2009 Consensus Conference on the management of adults with congenital heart disease: executive summary. Can J Cardiol 2010;26:143–50.

33. Marelli, A.J., Therrien, J., Mackie, A.S., Ionescu-Ittu, R., Pilote, L. (2009). Planning the specialized care of adult congenital heart disease patients; from numbers to guidelines; an epidemiologic approach. *Americal Heart Journal, 157,* 1-8.

34. Moons, P., Engelfriet, P., Kaemmerer, H., Meijboom, F.J., Oechslin, E., Mulder, B.J.M. (2006). Delivery of care for adult patients with congenital heart disease in Europe: results from the Euro Heart Survey. *European Heart Journal, 27,* 1324-1330.

35. de Bono, J., Freeman, L.J. (2005). Aortic coarctation repair – lost and found: The role of local long term specialized care. *International Journal of Cardiology, 104*, 176-183.

36. Kaemmerer, M., Vigl, M., Seifert-Klauss, V., Nagdyman, N., Bauer, U., Schneider, K.M., Kaemmerer, H. (2012). Counseling reproductive health issues in women with congenital heart disease. *Clinical Research Cardiology, 101,* 901-907.

37. Grattan, M., Power, A., Fruitman, D.S., Islam, S., Mackie, A.S. The impact of infective endocarditis prophylaxis recommendations on the practices of pediatric and adult congenital cardiologists. (2015). *Canadian Journal of Cardiology, 31,* e1497.e23-1497.e28.

38. Karsenty, C., Maury, P., Blot-Souletie, N., Ladoucer, M., Leobon, B., Senac, V., …, Hascoet, S. (2015). The medical history of adults with complex congenital heart disease affects their social development and professional activity. *Archives of Cardiovascular Disease, 108*(11), 589-597.

39. Sluman, M.A., Zomer, A.C., Vaartjes, I., Bouma, B.J., Mulder, B.J.M. (2014). Congenital heart disease may hurt men more than women in job participation. *International Journal of Cardiology, 172*(1): 230-232.

40. Zomer, A.C., Vaartjes, I., Uiterwaal, C.S.P., van der Velde, E.T., Sieswerda, G.T., Walon, E.M.C., …, Mulder, B.J.M. (2012). Social burden and lifestyle in adults with congenital heart disease. *American Journal of Cardiology, 109*, 1657-1663.

41. Kahr, P.C., Radke, R.M., Orwat, S., Baumgartner, H., Diller, G. (2015). Analysis of associations between congenital heart defect complexity and health-related quality of life using a meta-analytic strategy. *International Journal of Cardiology, 199*, 197-203.

42. White, K.S., Purdue, C., Ludbrook, P., Sodhi, S., Esmaeeli, A., Cedars, A. (2016). Cardiac denial and psychological predictors of cardiac care adherence in adults with congenital heart disease. *Behavior Modification, 40*(1-2), 29-50.

43. Khan, M., Monaghan, M., Klein, N., Ruiz, G., John, A.S. (2015). Associations among depression symptoms with alcohol and smoking tobacco use in adult patients with congenital heart disease. *Congenital Heart Disease, 10*, E243-E249.

44. Reid, G.J., Webb, G.D., McCrindle, B.W., Irvine, M.J., Siu, S.C. (2008). Health behaviors among adolescents and young adults with congenital heart disease. *Congenital Heart Disease, 3*, 16-25.

45. Clarizia, N.A., Chahal, N., Manlhiot, C., Kilburn, J., Redington, A.N., McCrindle, B.W. (2009). Transition to adult health care for adolescents and young adults with congenital heart disease: Perspectives of the patient, parent and health care provider. *Canadian Journal of Cardiology, 25*(9): e317-e322.

46. Bruce, E., Sundin, K. (2012). Experience of support for parents of adolescents with heart defects - supported to be supportive. *Journal of Pediatric Nursing*, *27*, 366-374.

47. Joesch, J.M., Smith, K.R. (1997). Children's health and their mothers' risk of divorce or separation. *Social Biology, 44*(3-4), 159-169.

48. Kovacs, A.H.., Utens, E.M. (2015). More than just the heart: transition and psychosocial issues in Adult Congenital Heart Disease. *Cardiology Clinics, 33,* 625-634.

49. Reid, G.J., Siu, S.C., McCrindle, B.W., Irvine, M.J., Webb, G.D. (2008). Sexual behavior and reproductive concerns among adolescents and young adults with congenital heart disease. *International Journal of Cardiology, 125*, 332-338.

50. Deng, L.X., Khan, A.M., Drajpuch, D., Fuller, S., Ludmir, J., Mascio, C.E., …, Kim, Y.Y. (2016). Prevalence and correlates of post-traumatic stress disorder in adults with congenital heart disease. *American Journal of Cardiology, 117,* 853-857.

51. Kovacs, A.H., Webb, G.D. (2015). Preparing pediatric patients for adult care: Are we ready? *The Journal of Pediatrics, 167*(6): 1194-1195.

52. Asp, A., Bratt, E., Bramhagen, A. (2015). Transfer to adult care - experiences of young adults with congenital heart disease. *Journal of Pediatric Nursing, 30*, e3-e10.

53. Van Deyk, K., Pelgrims, E., Troost, E., Goossens, E., Budts, W., Gewillig, M., et al. (2010). Adolescents' understanding of their congenital heart disease on transfer to adult-focused care. *American Journal of Cardiology, 106,* 1803-1807.

54. Goossens, E., Fieuws, S., Van Deyk, K., Luyckx, K., Gewillig, M., Budts, W., Moons, P. (2015). Effectiveness of structured education on knowledge and health behaviors in patients with congenital heart disease. *The Journal of Pediatrics, 166*(6), 1370-1376.

55. Jackson, J.J., Tierney, K., Daniels, C.J., Vannatta, K. (2015). Disease knowledge, perceived risk, and health behavior engagement among adolescents and adults with congenital heart disease. *Heart & Lung, 44*, 39-44.

56. Kantoch, M., Collins-Nakai, R., Medwid, S., Ungstad, E., & Taylor, D. (1997). Adult patients' knowledge about their congenital heart disease. *Canadian Journal of Cardiology, 13*(7), 641-645.

57. Bulat, D.& Kantoch, M. (2003). How much do parents know about their children's heart condition and prophylaxis against endocarditis? *Canadian Journal of Cardiology, 19*(5), 501-506.

58. Uzark, K., Smith, C., Donohue, J., Yu, S., Afton, K., Norris, M., Cotts, T. (2015). Assessment of transition readiness in adolescents and young adults with heart disease. *The Journal of Pediatrics, 167*, 1233-1238.

59. Reid, G.J., Webb, G.D., Barzel, M., McCrindle, B.W., Irvine, M.J., Siu, S.C. (2006). Estimates of life expectancy by adolescents and young adults with congenital heart disease. *Journal of the American College of Cardiology, 48*(2), 349-355.

60. Moons, P., Pinxten, S., Dedroog, D., Van Deyk, K., Gewillig, M., Hilderson, D., Budts, W. (2009). Expectations and experiences of adolescents with congenital heart disease on being transferred from pediatric cardiology to an adult congenital heart disease program. *Journal of Adolescent Health, 44,* 316-322.

61. Asp, A., Bratt, E., Bramhagen, A. (2015). Transfer to adult care - experiences of young adults with congenital heart disease. *Journal of Pediatric Nursing, 30*, e3-e10.

62. Lopez, K.N., Karlsten, M., De Nigris, F.B., King, J., Salciccioli, K., Jiang, A., …, Thompson, D. (2015). Understanding age-based transition needs: Perspectives from adolescents and adults with congenital heart disease. *Congenital Heart Disease, 10*(6): 561-571.

63. Bratt, E., Luyckx, K., Goossens, E., Budts, W., Moons, P. (2015). Patient-reported health in young people with congenital heart disease transitioning to adulthood. *Journal of Adolescent Health, 57,* 658-665.

64. Moceri, P., Goossens, E., Hascoet, S., Checler, C., Bonello, B., Ferrari, E., …, Fraisse, A. (2015). From adolescents to adults with congenital heart disease: the role of transition. *European Journal of Pediatrics, 174,* 847-854.

65. Heery, E., Sheehan, A.M., While, A.E., Coyne, I. (2015). Experiences and outcomes of transition from pediatric to adult health care services for young people with congenital heart disease: A systematic review. *Congenital Heart Disease, 10*, 413-427.

66. Hilderson, D., Saidi, A.W. Ven Deyk, K., Verstappen, A., Kovacks, A.H., Fernandes, S.M., …, Moons, P. (2009). Attitude toward and current practice of transfer and transition of adolescents with congenital heart disease in the United States of America and Europe. *Pediatric Cardiology, 30*, 786-793.

67. Valente, A.M., Landzberg, M.J., Gianola, A., Harmon, A.J., Cook, S., Ting, J.G., …, Gurvitz, M. (2013). Improving heart disease knowledge and research participation in adults with congenital heart disease (The Health, Education and Access Research Trial:HEART-ACHD). *International Journal of Cardiology, 168,* 3236-3240.

68. Chu, P.Y., Maslow, G.R., von Isenburg, M., Chung, R.J. (2015). Systematic review of the impact of transition interventions for adolescents with chronic illness on transfer from pediatric to adult healthcare. *Journal of Pediatric Nursing, 30*, e19-e27.

69. Okumura, M.J., Saunders, M., Rehm, R.S. (2015). The role of health advocacy in transitions from pediatric to adult care for children with special health care needs: Bridging families, provider and community services. *Journal of Pediatric Nursing, 30,* 714-723.

70. Walsh, G.W., Bondy, S.J., Rehm, J. (1998). Review of Canadian low-risk drinking guidelines and their effectiveness. *Canadian Journal of Public Health, 89*(4), 241-247.

71. Nishimura, R.A., Otto, C.M., Bonow, R.O., Ruiz, C.E., Carabello, B.E., Skubas, N.J., …, Thomas, J.D. (2014). AHA/ACC Guideline for the Management of Patients With Valvular Heart Disease: Executive Summary. *Circulation,* CIR.0000000000000031.

72. Mitchell, P., Powell, L., Blumenthal, J., Norten, J., Ironson, G., Pitula, C., et al. (2003). A short social support measure for patients recovering from myocardial infarction: The ENRICHD social support inventory. *Journal of Cardiopulmonary Rehabilitation, 23*, 398-403.

73. Reference: Spitzer, R., Kroenke, K., Williams, J., and the PHQ Primary Care Study Group. (1999). Validation and utility of a self-report version of the PRIME-MD: The PHQ primary care study. *Journal of the American Medical Association, 282*(18), 1737-1744.

74. Keir, M., Bailey, B., Lee, A., Kovacs, A.H., Roche, S.L. (2018). Narrative analysis of adults with complex congenital heart disease: childhood experiences and their lifelong reverberations. *Congenital Heart Disease, in press.*

75. Berkowitz, S.A., Fabreau, G.E. (2015). Food insecurity: what is the clinician's role. *Canadian Journal of Cardiology, 187*(14): 1031-1032.

76. Statistics Canada. https://www12.statcan.gc.ca/census-recensement/2016/dp-pd/prof/details/page.cfm?Lang=E&Geo1=PR&Code1=47&Geo2=PR&Code2=01&Data=Count&SearchText=Saskatchewan&SearchType=Begins&SearchPR=01&B1=Ethnic%20origin&TABID=1

77. Statistics Canada. Canadian Alcohol and Drug Use Survey (CADMUS) 2012. https://www.canada.ca/en/health-canada/services/health-concerns/drug-prevention-treatment/drug-alcohol-use-statistics/canadian-alcohol-drug-use-monitoring-survey-summary-results-2012.html#s9

78. Garne, E., Smaerup Olson, S., Paaske Johnson, S., Hjordtal, V., Orbaek Anderson, H., Nissen, H., …, Videbaek, J. (2012). How do we define congenital heart disease for scientific studies? *Congenital Heart Disease, 7*, 46-49.

79. Dash, J., Haller, D.M., Sommer, J., Junod Perron, N. (2016). Use of email, cell phone and text message between patients and primary-care physicians: cross-sectional study in a French-speaking part of Switzerland. *BMC Health Services Research*,*16*, 549 -555.

8.0 Appendices

Appendix A. Patient Telephone-Administered Survey

1. Patient Identification Number: _____

2. Birth year: _____ Age at Survey: _____

3. a) Were you born in Canada? ○ Yes ○ No
   b) If no, what year did you arrive in Canada? _____ Where from? _____

4. What language are you most comfortable communicating in?
_____

5. a) Do you feel comfortable talking to health care providers in English?
○ Yes ○ No
b)If no, have you ever had an interpreter? _____ Who? _____

6. How do you best define your ethnic group? _____

7. a) Do you have any of the following disabilities?
○ Learning Disability ○ Physical Disability ○ Hearing Disabiity
○ Visual Disability ○ Other disability

b) If yes to above, what is the nature of your disability?
_____

9. What is the highest educational level you completed:
○ <high school
○ completed high school
○ some college/university
○ college/university degree
○ advanced degree

10. Which of the following best describes your current housing situation?
○ renting
○ home you own

O home owned by parents/guardian

O group home/boarding home/supportive housing

O homeless/on the street/shelter

O correctional facility

11. Regarding the food situation in your household during the past 12 months, how often was the following statement true?

The food that you and other household members bought just didn't last and there wasn't any money to buy more.

O Never true O Sometimes true O Often true

12. When you were growing up, who did you live with?

With a biological parent: O single parent household O two parent household

With adopted parent: O single parent household O two parent household

O With a family member: _____

O In foster care/group home

13a). What is your family income per year?

O < $25,000

O $25,000 - $50,000

O $50,000 – $100,000

O >$100,000

b) How many people does that income support? _____

14. Are you married or do you have a common-law spouse? O yes O no

15. Do you?

O work full-time for pay

O work part-time for pay

O not currently work for pay

16. a) If you work for pay, what is your occupation or job?

_____

b) Have you ever been unemployed because of your congenital heart disease?

_____

_____

17. Do you smoke cigarettes? O yes O no

b) If no, have you ever smoked cigarettes? O yes O no

c) If yes, when did you start and when did you quit? _____
How many cigarettes a day did you smoke, on average? _____

18. a) Do you drink alcohol? O yes O no

b) If yes, how many drinks per week, on average?

_____

19. a) Have you ever done drugs that were not prescribed by a physician?

O yes O no

b) If yes, which ones?

_____

20. What is the medical name for your heart condition?

_____

21. In your own words, can you tell me what is wrong with your heart?

_____

_____

_____

_____

24. Have you had any heart operations? If so, could you tell me the medical name of the operations?

_____

_____

_____

_____

25.a) Do you take any heart medications?

O yes O no

b) If yes, what are the names of your medications and what do they do?

Name of Medication	Reason you take it

c) If you are biologically female, are you using a form of contraception?

O yes O no

If yes, what type: _____

26a). Do you have a family physician?

O yes O no

b) How often do you see your family physician, on average?

O every 6 months O every year O every two years O every 3-5 years

27. Do you think you need to see an adult congenital heart disease specialist doctor regularly?

O yes O no

28. Who is/are the doctor(s) that manage your heart condition?

_____

29. When is the last time you saw this doctor about your heart? _____

30. How often do you have appointments to see your heart doctor?

O every 6 months O every year O every two years O every 3-5 years

31. a) Have you ever missed an appointment with your heart doctor? O yes O no

b) If yes, why?

_____

_____

_____
_____

32. When you were a teenager (middle adolescence (age 15-18)), did you ever attend cardiology appointments without a parent/guardian or sibling?

O yes O no

33.a) If biologically female, have you ever been pregnant? O yes O no

b) If no, has a doctor ever told you the risks to your heart from pregnancy?

O yes O no

c) If yes, did a doctor discuss the risks of pregnancy to your heart before you got pregnant?

O yes O no

d) If yes, did a doctor discuss the risk of congenital heart disease to your baby and recommend a special fetal screening ultrasound?

O yes O no

e) If yes, what was the outcome(s) of the pregnancy?

_____
_____
_____

f) If yes, did you see a heart doctor during your pregnancy? O yes O no

g) Did you have any medical problems/complications during your pregnancy?

_____
_____
_____

34. If yes to 33a) or biologically male, do you have any children (living or deceased)?

O yes O no

b) If yes, how many? _____

c) If yes, do they have any heart problems?

_____

_____
_____

35.a) Do you take antibiotic medicine before dental procedures?

○ yes ○ no

b) If no, has a doctor every told you that you should take antibiotic medicine before dental work?

○ yes ○ no

c) Why do patients with heart defects need to take medicine before dental work?

_____
_____
_____

d) If yes to a), can you think of any situation (other than before dental work) that you would need to take antibiotic medicine?

_____
_____
_____
_____

36. Perform ENRICHD Social Support Survey (**see Appendix B**)

Low Perceived Social Support (LPSS)? ○ yes ○ no

37. Perform Krantz HOS Survey (**see Appendix C**)

Score: ○ high ○ medium ○ low

38. Perform Depression Screen (**see Appendix D)**

Major Depressive Disorder: ○ yes ○ no

Appendix B. ENRICHD Social Support Instrument (ESSI)

Please listen to the following questions and tell me which response most closely describes your current situation?

1. Is there someone available to you whom you can count on to listen to you when you need to talk?

○ none of the time(1) ○ a little of the time(2) ○ some of the time(3)

○ most of the time(4) ○ all of the time(5)

2. Is there someone available to give you good advice about a problem?

○ none of the time(1) ○ a little of the time(2) ○ some of the time(3)

○ most of the time(4) ○ all of the time(5)

3. Is there someone available to you who shows you love and affection?

○ none of the time(1) ○ a little of the time(2) ○ some of the time(3)

○ most of the time(4) ○ all of the time(5)

4. Is there someone available to you to help with daily chores?

○ none of the time(1) ○ a little of the time(2) ○ some of the time(3)

○ most of the time(4) ○ all of the time(5)

5. Can you count on anyone to provide you with emotional support (talking over problems or helping you make a difficult decision)?

○ none of the time(1) ○ a little of the time(2) ○ some of the time(3)

○ most of the time(4) ○ all of the time(5)

6. Do you have as much contact as you would like with someone you feel close to, someone in whom you can trust and confide?

○ none of the time(1) ○ a little of the time(2) ○ some of the time(3)

O most of the time(4) O all of the time(5)

7. Married or living with a partner? NOTE: Data already collected …. Do not ask question …

O yes(4) O no(2)

Score: less than 3 on ≥ 2 items and a total score of less than 18, or less than 2 on ≥ 2 items = low perceived social support (LPSS)

References:

Mitchell, P., Powell, L., Blumenthal, J., Norten, J., Ironson, G., Pitula, C., et al. (2003). A short social support measure for patients recovering from myocardial infarction: The ENRICHD social support inventory. *Journal of Cardiopulmonary Rehabilitation, 23*, 398-403.

Writing Committee for the ENRICHD Investigators. (2003). Effects of treating depression and low perceived social support on clinical events after myocardial infarction: The enhanced recovery in coronary heart disease patients (ENRICHD) randomized trial. *Journal of the American Medical Association, 289*(23), 3106-3116.

Appendix C. Krantz Health Opinion Survey (HOS)

1. I usually don't ask the doctor or nurse many questions about what they're doing during a medical exam. (I)

○ agree (0) ○ disagree (1)

2. Except for serious illness, it's generally better to take care of your own health than to seek professional help. (B)

○ agree (1) ○ disagree (0)

3. I'd rather have doctors and nurses make the decisions about what's best than for them to give me a whole lot of choices. (I)

○ agree (0) ○ disagree (1)

4. Instead of waiting for them to tell me, I usually ask the doctor or nurse immediately after an exam about my health. (I)

○ agree (1) ○ disagree (0)

5. It is better to rely on the judgement of doctors (who are the experts) than to rely on "common sense" in taking care of your own body. (B)

○ agree (0) ○ disagree (1)

6. Clinics and hospitals are good places to go for help since it's best for medical experts to take responsibility for health care. (B)

○ agree (0) ○ disagree (1)

7. Learning how to cure some of your own illness without contacting a physician is a good idea. (B)

○ agree (1) ○ disagree (0)

8. I usually ask the doctor or nurse lots of questions about the procedures during a medical exam. (I)

○ agree (1) ○ disagree (0)

9. It's almost always better to seek professional help than to try to treat yourself. (B)

○ agree (0) ○ disagree (1)

10. It is better to trust the doctor or nurse in charge of a medical procedure than to question what they are doing. (I)

○ agree (0) ○ disagree (1)

11. Learning how to cure some of your illness without contacting a physician may create more harm than good. (B)

○ agree (0) ○ disagree (1)

12. Recovery is usually quicker under the care of a doctor or nurse than when patients take care of themselves. (B)

○ agree (0) ○ disagree (1)

13. If it costs the same, I'd rather have a doctor or nurse give me treatments than to do the same treatments myself. (B)

○ agree (0) ○ disagree (1)

14. It is better to rely less on physicians and more on your own common sense when it comes to caring for your body. (B)

○ agree (1) ○ disagree (0)

15. I usually wait for the doctor or nurse to tell me the results of a medical exam rather than asking them immediately. (I)

○ agree (0) ○ disagree (1)

16. I'd rather be given many choices about what's best for my health than to have the doctor make the decisions for me. (I)

agree (1) disagree (0)

Low Score = 6 or less

Medium Score = 7 - 9

High Score = 10 - 16

Reference: Krantz, D., Baum, A., Wideman, M. (1980) Assessment of preferences for self-treatment and information in health care. Journal of Personality and Social Psychology, 39(5): 977-990.

Appendix D. PHQ-9 Survey for Depression Screening

Over the last two weeks, how often have you been bothered by any of the following problems?

	Not at all	Several days	More than half the days	Nearly every day
1. Little interest or pleasure in doing things	0	1	2	3
2. Feeling down, depressed, or hopeless	0	1	2	3
3. Trouble falling or staying asleep, or sleeping too much	0	1	2	3
4. Feeling tired or having little energy	0	1	2	3
5. Poor appetite or overeating	0	1	2	3
6. Feeling bad about yourself – or that you are a failure or have let yourself or your family down	0	1	2	3
7. Trouble concentrating on things, such as reading the newspaper or watching television	0	1	2	3
8. Moving or speaking so slowly that other people have noticed. Or the opposite – being so fidgety or restless that you have been moving around a lot more than usual	0	1	2	3
9. Thoughts that you would be better off dead, or of hurting yourself	0	1	2	3

Scoring: Score of 10 or greater and a score of 2 or more on either Q1 or Q2 indicates major depressive disorder.

Reference: Spitzer, R., Kroenke, K., Williams, J., and the PHQ Primary Care Study Group. (1999). Validation and utility of a self-report version of the PRIME-MD: The PHQ primary care study. *Journal of the American Medical Association, 282*(18), 1737-1744.

Appendix E. Data Collection Form: Patient Chart Review

Patient Identification Number: _____

Biological Sex: ○ Male ○ Female

Year of Birth: _____

Date of Transition: _____

Age at Transition: _____ years _____ months

Congenital Heart Lesion & Procedures Performed (Surgical and/or Interventional):
_____
_____
_____
_____

Grade of Lesion (see **Appendix F**):

○ Great Complexity ○ Moderate Complexity ○ Simple

Distance of Transition Address from Tertiary Care Centre: _____ km

Closest Tertiary Care Centre:

○ Saskatoon ○ Regina ○ Edmonton ○ Winnipeg

Level of Specialist Care (**see Appendix G**):

○ 1 ○ 2 ○ 3

Level of Care Appropriate for Lesion Severity (**see Appendix H**)? : ○ yes ○ no

Date of Last Appointment Since Transition: _____

Average Rate of Follow-Up:

○ every 6 months ○ every year ○ every two years ○ every 3-5 years

Appropriate Rate of Follow-up as a child? (**see Appendix H**): ○ yes ○ no

Appropriate Rate of Follow-up (**see Appendix H**): ○ yes ○ no

Missed Appointments: ○ yes ○ no

Number of missed appointments since transition: _____

Medical Comorbidities: ○ yes ○ no

If yes, please list:

_____
_____
_____
_____

Cardiac Complications Since Transition?

Arrhythmia: ○ yes ○ no

If yes, what?: _____
Date: _____
Treatment: _____

Hospitalizations: ○ yes ○ no

If yes, why? _____
Dates of admission: _____
Treatments: _____

Endocarditis: ○ yes ○ no

If yes, when? _____
Lesion & organism? _____
Treatment: _____

Repeat Cardiac Surgery/Intervention: ○ yes ○ no

If yes, what surgery? _____
When? _____

Requires Dental Prophylaxis (**see Appendix I**): ○ yes ○ no

Appendix F. Grading of Congenital Heart Lesion Severity

Complexity	Diagnosis
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
Moderate Complexity	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

Simple	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

Reference: Warnes, C., Liberthson, R., Danielson, G., Dore, A., Harris, L, Hoffman, J., …, Webb.G. (2001). Task force 1: The changing profile of congenital heart disease in adult life. *Journal of the American College of Cardiology, 37*(5), 1170-1175.

Appendix G. Appropriate Level of Specialist Care for ACHD Patients

Level of Care	Definition
1 = Specialist	Follow-up given by specialized ACHD cardiologist at a tertiary care center
2 = Shared Care	Follow-up given by a general adult cardiologist in close collaboration with a congenital heart disease specialist
3 = Nonspecialist Care	Follow-up given by a general or community cardiologist, or a general practitioner, with access to specialized care if needed

Reference: Deanfield, J., Thaulow, E., Warnes, C., Webb, G., Kolbel, F., Hoffman, A., …, Trappe, H. Management of grown up congenital heart disease: The task force on the management of grown up congenital heart disease of the European Society of Cardiology. *European Heart Journal* (2003): 24, 1035-1084.

Appendix H. Appropriate Follow-Up for ACHD Patients

Risk Category	Appropriate Follow-Up
Simple	Every 3-5 years, Nonspecialist Care
Moderate Complexity	Every 1-2 years, Shared Care
Great Complexity	Every 1-2 years, Specialist Care
Great Complexity with: single ventricle, morphological RV in systemic circuit, heart failure, recurrent arrhythmias, or pulmonary vascular obstructive disease	Every 6-12 months, Specialist Care

Reference: Landzberg, M., Murphy, D., Davidson, W., Jarco, J., Krumholz, H., Mayer, J., …, Williams, R. (2001). Task force 4: Organization and delivery systems for adults with congenital heart disease. *Journal of the American College of Cardiology, 37*(5), 1187-1193.

Appendix I. Appropriate Endocarditis Prophylaxis Based on Lesion/Patient Characteristics

Patients with the Highest Risk of Developing Endocarditis after Invasive Dental Procedures: Indications for Prophylaxis
1) Previous Infective Endocarditis
2) Prosthetic cardiac valve or prosthetic material used for cardiac valve repair
3) Unrepaired cyanotic CHD, including palliative shunts and conduits
4) Completely repaired CHD with prosthetic material or device, whether by surgery or by catheter intervention, during the first 6 months of the procedure
5) Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device that inhibit endothelialization
6) Cardiac transplant recipients who develop cardiac valvulopathy

Reference: Warnes, C. A., Williams, R. G., Bashore, T. M., Child, J. S., Connolly, H. M., Dearani, J. A., et al. (2008). ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: A report of the american college of Cardiology/American heart association task force on practice guidelines (writing committee to develop guidelines on the management of adults with congenital heart disease): Developed in collaboration with the american society of echocardiography, heart rhythm society, international society for adult congenital heart disease, society for cardiovascular angiography and interventions, and society of thoracic surgeons *Circulation, 118*(23), 714-833.