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Ensuring a successful transition and transfer from pediatric to adult care in patients with congenital heart disease

As CHD patients make the shift from a family-centred pediatric model of care to an autonomous adult model, self-management and self-advocacy skills become essential, and resources available through the Transitioning Responsibly to Adult Care initiative (ON TRAC) can be useful.

ABSTRACT: Patients with congenital heart disease require lifelong surveillance for arrhythmias, ventricular failure, and complications associated with childhood surgery. When pediatric patients with congenital heart disease become adults they can be lost to follow-up or experience lapses in care that expose them to greater risk of adverse health outcomes. A successful transition and transfer from pediatric to adult care supports youth attachment to both a primary care provider and a specialized cardiology clinic—ensuring quality and continuity of care. Transition refers to a process that begins in early adolescence and continues through early adulthood when health care management shifts from the family to the patient. Transfer is an event that occurs when the responsibility for patient care moves from one health care team to another. In BC pediatric patients with congenital heart disease are transferred from the Children's Heart Centre to the Pacific Adult Congenital Heart Disease program at age 18. Resources that can aid in transition and transfer include toolkits developed through the Transitioning Responsibly to Adult Care initiative and information provided by the iHeartChange website. Primary care considerations for young adult patients include cardiac surveillance and screening, sexual and reproductive health, and psychosocial health. The population of congenital heart disease patients is growing and aging, and continued attention will be needed to ensure these patients move successfully from pediatric to adult care.

This article has been peer reviewed.

Population estimates indicate there are approximately 12 000 adults with moderate and severe congenital heart disease (CHD) in British Columbia. The significant increase in patient numbers (estimated growth to be more than 5% per year) is attributable to the excellent survival outcomes for young patients with CHD achieved in past decades and the greater awareness of services for these patients. Services include pediatric care provided through the Children's Heart Centre at BC Children's Hospital (BCCH) and adult care provided through the Pacific Adult Congenital Heart Disease (PACH) clinic at St.

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Paul's Hospital. Ensuring a successful transition and transfer from pediatric to adult care is critical to maintaining patient health in adulthood.

There are 15 specialized adult CHD centres across the country, all belonging to the Canadian Adult Congenital Heart network or CACH network (www.cachnet.org). The PACH clinic is one of five that offer a full range of services and qualifies as a supraregional centre. CACH network has been instrumental in establishing templates and guidelines for standardized follow-up of many adult CHD conditions. Building on this work, the Canadian Cardiac Society (CCS) convened a panel of international experts for an adult CHD consensus conference in 1996.[1] Foremost among the proposals arising from this forum was a recommendation that all patients be referred to specialized centres for ongoing care. Mylotte and colleagues documented an increase in referrals to specialized centres in Quebec after publication of the CCS recommendations in 1998, and demonstrated that referral was independently associated with a significant decrease in mortality, supporting the model of specialized care for patients with adult CHD.[2]

Pacific Adult Congenital Heart clinic

The PACH clinic was established at Shaughnessy Hospital in 1988 to respond to a gap in health services for adults with CHD, and moved to St. Paul's Hospital in 1993. The PACH team, a multidisciplinary group of health professionals with advanced training in adult CHD, is a unique provincial resource that provides comprehensive health services to CHD patients. The core team includes adult and pediatric cardiologists, cardiac surgeons, cardiac radiologists, patient educators (registered nurses), a clinical nurse specialist, a psychologist, a genetic counselor, and a social worker. Additional specialists are recruited as required. Services provided include consultation, ongoing medical care, cardiac surgery, cardiac intervention, electrophysiology procedures, diagnostic imaging, obstetrical care, and patient and family education and counseling.

A team approach to care is integral to optimizing the health of this complex patient population.

Patients are discussed at weekly case conferences and bimonthly morbidity and mortality rounds. Cardiology fellows, residents, medical students, and other health care professionals in training also attend, which ensures ongoing education for those caring for this patient population. There is a PACH physician on call 24 hours a day, 7 days a week, to assist physicians and other health care providers throughout the province. The PACH team provides comprehensive, cost-effective care to adults with CHD.

A team approach to care is integral to optimizing the health of this complex patient population. The PACH patient educators are the initial point of contact for both patients and community health care providers. The patient educators provide case management and liaise on a daily basis with the PACH team to support patient wellness and assist with the challenges of navigating the health care system. During clinic visits, patient-centred education is based on a chronic disease self-management model. When patients have a better understanding of their individual cardiac condition they are able to recognize concerning symptoms, communicate effectively with their health care providers, and determine when it is appropriate to seek medical attention. Patient educators are available to provide ongoing support in these areas and to facilitate self-management by connecting patients with resources when necessary. Telehealth services support patients and their health care providers to manage their CHD and facilitate additional patient education outside clinic hours.

The PACH clinic currently follows 2700 active patients and has a wait time of 3 to 4 months for patients to be seen on a nonurgent basis. The volume of patients served has almost tripled in the last decade and the clinic has been advocating for expanded services. To accommodate the growing population of adults with CHD throughout BC, a closer-to-home care model, patterned on the success

of BCCH's Cardiology Partnership Program, is being explored to bring adult CHD services to communities outside the Lower Mainland. Partnering with communities and local health care providers has proven successful when serving other populations with chronic health conditions. Bringing the expertise of adult CHD specialists to communities across the province would support care providers in local communities to deliver quality care to this group of complex patients.

Moving from pediatric to adult care

Currently most children born with CHD are surviving, and can be expected to thrive well into their adult years. There are now more adults than children with CHD. With the surgical advancements of the past several decades, 95% of pediatric congenital heart disease patients will now transfer to adult care, the largest growth in this population being youth with complex CHD.[3,4] Adults with CHD are pursuing advanced education, building careers, and starting families of their own. Lifelong cardiac follow-up is required to achieve the best health outcomes for these patients. To support seamless cardiac care across the lifespan of a congenital heart patient, a structured transition process followed by a transfer of care is necessary.

Transition and transfer

A successful transition and transfer from pediatric to adult CHD care supports youth attachment to a primary care physician and specialty clinic(s) and ensures continuity of care. Transition refers to a process that begins in early adolescence and continues through early adulthood when health care management shifts from the family to the patient.[5,6] Transfer is an event that occurs when the patient moves from one health care team to another. When transition and transfer from pediatric to adult health care are less than optimal, the repercussions can be serious, and may include hospital admissions, adverse health events, and even death.[7] The potential for residual disease and complications following childhood management of CHD necessitates lifelong surveillance for arrhythmias, ventricular failure, and the need for further surgery.[3]

An estimated 30% to 70% of CHD patients are lost to follow-up or experience lapses in care,[8,9] situations that expose them to greater risk of adverse health outcomes. Gaps in care are more frequent in males, in those with mild and moderate defects, and in patients with a history of follow-up outside an adult CHD clinic.[4,8,9] Additional challenges to successful transition include patient adherence, relocation, and attachment to a specialty clinic.[6] As CHD patients move from a family-centred pediatric model of care to an autonomous adult model, self-management and self-advocacy skills become essential.[10] Transition and transfer must be seen as essential elements in the care of patients with CHD. Expert consensus recommends that structured plans be developed to facilitate the transition process.[6]

Multiple tools and strategies have been developed and are being evaluated to support the transition and transfer process for youth with CHD as they move from the Children's Heart Centre at BCCH to the PACH program at St. Paul's Hospital when they reach age 18.

ON TRAC initiative

Transitioning Responsibly to Adult Care (ON TRAC) is a provincial initiative that supports youth with chronic health conditions as they prepare to move from pediatric care and then as they transfer and attach themselves to the adult health care system. Using a framework from the Institute for Healthcare Improvement Triple Aim framework, ON TRAC consists of four separately funded projects focused on policy change, clinical practice, health system performance, and youth engagement. Key stakeholders are involved at every level to inform decisions, develop and test tools, and shape recommendations. ON TRAC is supported with funding from the Vancouver Foundation, BCCH, Child

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Health BC, and the Shared Care Committee (SCC) and Specialist Services Committee (SSC)—joint committees of Doctors of BC and the BC Ministry of Health.

ON TRAC reflects work done by groups of pediatric and adult care providers, patients, and families to address gaps that have traditionally been a challenge in the transition from pediatric to adult care by:

- Preparing youth and families with skills to function in the adult system.
- Supporting health care providers through clinical guidelines, training, tools, and online resources.
- Defining the role of community-based family physicians in providing continuity of care.
- Defining appropriate referral pathways and care requirements for youth with complex health conditions.

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The ON TRAC package of transition and transfer tools for CHD patients, family members, and care providers was developed to ensure that a comprehensive transfer of care occurs when the transition is complete. The package includes:

- Transition Clinical Pathways (TCP) form: Developed through BCCH, this form is used to guide the preparation of youth and document their readiness to manage their care in the adult care system. The TCP is provided to adult specialists and community-based family practitioners at the time of transfer. The form lists cardiac-specific tests and reports and highlights areas where additional education and support are required to help the young patient acquire the knowledge and skills needed for self-management. (www.bcchildrens.ca/transition-to-adult-care/Documents/TCPCOMPLEXCARDIOLOGY.pdf)
- Medical Transfer Summary (MTS) form: Developed with the support of BCCH and the SCC, this form ensures that the family physician, community care providers, and the specialty clinic receive a comprehensive summary of the medical history (including details about investigations, surgeries, medications, immunizations), psychosocial considerations and anticipatory guidance and recommendations for future care. (www.bcchildrens.ca/transition-to-adult-care/Documents/MTSCARDIOTemplateDec2015.pdf)
- Transition Care Management Plan (TCMP): Developed with the support of the SSC, the TCMP helps community care providers understand the care requirements of patients with CHD. The TCMP consists of background information (purpose, brief description of lesion and management, recommendations for use of the plan) and delineates the role of the primary care physician and adult CHD specialist. It outlines the potential risks associated with the specific lesion, health surveillance and clinical evaluation recommendations, sexual and reproductive health considerations, and noncardiac surgery and procedures. The TCMP also outlines patient counseling recommendations for medications, exercise, and lifestyle considerations. TCMPs will be available online beginning November 2016. The TCMP website will contain the template and tips on how to develop a plan. These materials will be posted on the ON TRAC website in the health care provider toolkit.
- ON TRAC website: The website provides access to toolkits for youth, families, and health care providers. The youth toolkit helps patients develop the skills and obtain the support required for an effective transition into adulthood and the adult health care system. With the help of videos, self-directed activities, and other resources, youth can develop self-advocacy and self-management skills, obtain peer support, engage in educational, vocational, and financial planning, and learn more about sexual health and healthy lifestyle choices. The family toolkit helps family members provide guidance regarding additional support for housing, fi-

nances, personal care, and guardianship. The health care provider toolkit includes templates, guidelines, and resources to assist those supporting youth through the transition process. (www.ontracbc.ca)

iHeartChange

The website iHeartChange (https://iheartchange.org) is a CHD-specific, empirically studied Internet resource developed for youth, families and friends, and health care providers. The website provides introductions to adult care teams in many North American communities, medical and lifestyle information, and suggestions for becoming independent and coping with CHD. Users must create an account to access the site, but there is no cost or restriction otherwise. Youth can earn a Transition Diploma by visiting the various sections of the site and answering questions about living with CHD.[11]

Primary care considerations

The primary care physician plays an essential role in the transition and transfer process. Children and youth who are not well connected to a primary care physician may not develop an effective relationship with the health care system as young adults.[12] Given the centralized nature of subspecialty care, young adult CHD patients may also not learn about the resources available in their home communities.[13] In addition, concerns have been raised by general internal medicine specialists regarding inadequate exposure and knowledge regarding children with complex pediatric disorders and the unique health and psychosocial aspects of these adolescents and young adults.[14]

Young adults who have a strong relationship with a community physician are more likely to have a successful transition to adult care and to receive appropriate health maintenance advice and ongoing care regarding the following primary care considerations.

Surveillance, screening, and counseling

Patients with CHD and acquired valvular heart disease are at risk for infective endocarditis and may require antibiotic prophylaxis.[15] As well as being considered for appropriate antibiotic prophylaxis, patients should be screened for acquired cardiac risk factors such as hypertension and hypercholesterolemia, and counseled regarding nutrition and the use of tobacco, marijuana, alcohol, and drugs. Lifestyle choices such as body piercing and tattoos should also be discussed.

Sexual and reproductive health

Discussion may be needed about concerns regarding body image that can affect a young patient's self-esteem and have an impact on sexuality and sexual choices. The risk of having a child with CHD should be reviewed with both men and women, and fetal echocardiography should be offered to all prospective parents. Specific issues regarding contraception and pregnancy must be discussed with women. Most women with CHD have no limits on contraceptive choice, and counseling can focus on efficacy and individual needs. The exception is women with an increased thrombosis risk in which the use of combined estrogen/progestin oral contraceptives should be avoided. This group can use progestin (only) agents, such as oral or implanted agents.

Preconception counseling to review the potential risks and safety of a pregnancy may be organized with the Cardiac Obstetrics (COB) clinic at St. Paul's Hospital.

Sports and physical activity participation

The vast majority of youth and young adults with CHD can participate in physical activity and sport with minimal restrictions. Exercise recommendations outlined by the congenital cardiologist should be supported by the primary care provider.

Psychosocial health

Health care providers may fail to detect psychosocial distress if they do not ask about it or deliberately screen for it as part of routine care. The most straightforward way is to ask patients about specific challenges or difficult feelings that they may be experiencing in daily life, and to provide appropriate mental health referrals when needed. In North America, approximately 1 in 3 adults with CHD have clinical levels of depression and anxiety,[16-19] even when these patients are assumed to be well adjusted by their cardiologists.[16] Regardless of formal psychiatric status, many adults with CHD face potential psychosocial challenges as a result of growing up with a complex health condition. Common challenges include disease management, intrapersonal and emotional issues, impaired social functioning, educational and vocational difficulties, and poor health behaviors.[20-23] It is unclear whether psychosocial functioning is worst among patients with more complex CHD.[23] A number of factors[24,25] may contribute to lower psychological functioning:

- Female sex.
- Low capacity for exercise.
- Restrictions placed by physician.
- Body image concerns/perception of scarring.
- Perceived health status.
- Loneliness/social anxiety/fear of negative evaluation/poor social support/poor social problem-solving abilities.

- Poor academic performance.
- Perceived financial strain.

Many patients with CHD report feeling "different," and the therapeutic relationship may improve if both patients and providers view themselves as partners of equal status in the health care process. There are a number of excellent review papers about common psychosocial issues in CHD for the interested reader.[23,25-27]

Referring patients to the PACH clinic

Patients age 18 and older with moderate and severe congenital heart disease (**Table**) should be referred to the PACH clinic (see **Box** for contact information). Patients with simple disease may benefit from visiting the clinic once to confirm that their diagnoses are correct. Patients with simple disease may also visit the clinic under special circumstances such as during pregnancy, or to obtain advice on treating complications such as arrhythmias.

Patients with an urgent referral can be seen within days. For nonurgent referrals, the wait time is approximately 3 to 4 months. Before an appointment is booked, the clinic obtains all previous cardiac catheterization and cardiac surgical reports. Each patient is unique and surgical repairs may have been modified. Reports from previous consultations and procedures (pediatric cardiology, cardiac surgery, and adult cardiology) are helpful, as are imaging reports. The clinic orders appropriate investigations prior to the patient's clinic appointment, usually on the same day for patients from outside the Lower Mainland, so that a comprehensive assessment can be provided at the time of the consultation.

Summary

Approximately 12 000 adults with moderate and severe congenital heart disease live in British Columbia. Pediatric patients with CHD are followed through the Children's Heart Centre at BCCH and are transferred to the PACH program at St. Paul's Hospital at age 18. To accommodate the growing population of adults with CHD, a closer-to-home care model is being explored as a way to bring adult congenital cardiology services to additional communities throughout BC.

A successful transition and transfer from pediatric to adult care supports youth attachment to a primary care provider and ensures that continuity of care is maintained. Resources that aid in transition and transfer include toolkits developed for the ON TRAC initiative and the iHeartChange website.

Primary care considerations include surveillance and screening, sexual and reproductive health, and psychosocial health. Patients with moderate and severe CHD should be followed by the multidisciplinary team at the PACH clinic, while those with simple disease may benefit from visiting the clinic once to confirm that their diagnoses are correct.

The population of adults with CHD is growing and aging. The success of pediatric care providers in achieving excellent survival outcomes for young patients requires that we maintain the momentum

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Table. Congenital heart defects by severity.

Simple	Moderate	Severe
• Isolated congenital valve disease • Small, isolated atrial or ventricular septal defect • Repaired atrial or ventricular septal defect • Repaired patent ductus arteriosus	• Atrioventricular canal defect • Anomalous pulmonary veins • Coarctation of the aorta • Ebstein anomaly • Ostium primum atrial septal defect • Sinus venosus atrial septal defect • Patent ductus arteriosus not closed • Right ventricular outflow tract obstruction • Moderate to severe pulmonary stenosis or insufficiency • Subvascular or supravalvular aortic stenosis • Tetralogy of Fallot • Ventricular septal defect with associated defects	• Obstructed conduit • Cyanotic heart disease • Double outlet right ventricle • Eisenmenger syndrome • Fontan procedure complication • Single-ventricle defect • Transposition of the great arteries • Truncus arteriosus • Isomerism • Heterotaxy syndrome

and ensure that adult CHD patients receive the quality and continuity of care they need.

Competing interests
None declared.

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