


# Historical investigation of medical treatment for adult congenital heart disease: A Canadian perspective

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## Abstract

**Objective:** The number of adults with congenital heart disease (CHD) has increased substantially because of medical advances that are extending life expectancy beyond childhood. A historical investigation is timely not only because of this significant demographic shift, but also because stakeholder engagement is increasingly important in shaping research directions. The study questions were: From the perspective of stakeholders, what is known about the medical treatment for the adult CHD cohort in Canada and how has it changed over time?

**Design/Methods:** We harnessed the use of historical research methods and conducted interviews with 21 key informants in order to articulate a contextualized understanding of the evolution of adult CHD treatment in Canada. The informants recruited were currently or previously located in and/or involved in treatment in seven provinces and two territories across Canada including patients, families, advocates, researchers, and practitioners located in disciplines that included cardiology, cardiovascular surgery, nursing, psychology, dietetics, and kinesiology.

**Results:** Alongside findings that highlighted the significant demographic shift, the findings highlight key themes related to temporal shifts in treatment, emergence of organizational structures and use of evidence, comprehensive approaches to care, and future directions. A critical finding that requires immediate attention is the significantly disproportionate resources to the number of adults living with CHD, and as a result, the real risk of premature death for this population.

**Conclusions:** The insights provided behoove the community of stakeholders to think creatively on how to draw attention to the inadequacy of resources and the unique and diverse needs of this population.

## KEYWORDS

adults, congenital heart disease, heart disease, historical methods, qualitative research

## 1 | INTRODUCTION

Adults with congenital heart disease (CHD) encompass a growing population who require lifelong care. Historically, CHD resulted in infant/childhood mortality in which adulthood was often not reached particularly with complex cases. Individuals are now increasingly reaching adulthood<sup>1-3</sup> with a dramatic increase of those over 65<sup>4</sup> as a result of advances in diagnostics, medical management, surgical interventions, and congenital cardiology field.<sup>4-6</sup> Empirical data indicated nearly two thirds of individuals with CHD were adults.<sup>7</sup>

A historical investigation is timely not only because of this significant demographic shift, but also because stakeholder engagement is increasingly important in shaping research directions.<sup>8</sup> This shift is a public health concern that impacts health service utilization and costs.<sup>7</sup> Adults with CHD have unique and complex needs such as but not limited to comorbidities, arrhythmias, mental health concerns, and increased cardiovascular risk factors and hospitalization rates. These individuals still encounter limitations in quality of life and longevity that are distinct from others with acquired heart disease, and a historical perspective may shed light on the trajectory of advances in the care of people with adult congenital heart disease (ACHD), and the areas in which further efforts are required.

Historical research approaches are of benefit because of the capacity to produce extensive accounts of history, explicate trends, and map strategic steps.<sup>9</sup> We harnessed the use of historical methods in order to articulate a contextualized understanding of the evolution of ACHD treatment in Canada. The study questions were: From the perspective of stakeholders/key informants, what is known about medical treatment for the ACHD cohort and how has it changed over time? A broad conception of medical treatment was assumed including the various health care services and care provided by interdisciplinary team members that address physical, psychological, and social dimensions of health. In this article, we describe the study methods and findings.

## 2 | METHODS

A historical research methodology was employed, which involves systematic examination and recapturing accounts of the past through oral histories with key informants.<sup>9</sup> This data source provides nuanced and complex information that is often carried through stories and not necessarily documented in written forms.<sup>9</sup> Using a responsive design, recruitment, interviewing, and analysis were informed by the unfolding history told through oral traditions. Semi-structured interview guides were employed so that informants' stories directed the flow of conversation based on their experiences. Interview guides were a living document, reshaped based on early data collection and thematic analysis as well as individualized to key informants. Using thematic analysis, the interview transcripts were reviewed in a group setting by 2-3 research team members. The team engaged in a dialogical process of justifying analytic decisions and arrived on consensus about the coding of data by engaging iterative discussions. A study report, highlighting the main themes with representative quotes, was shared with the investigative team ( $n = 3$ ) and the advisory committee ( $n = 3$ ), as well as key informants ( $n = 16$ ) who indicated on the consent form that they were interested in receiving this report.

Informants were recruited using purposive and snowball sampling.<sup>10,11</sup> The research team identified information-rich informants based on the project goal and diversity of experiences over varying time periods. Informants were also asked to identify individuals who they believed could provide important information about the topic.<sup>11</sup> Either currently or in the past, the 21 informants recruited were located in and/or involved in treatment in 7 provinces and 2 territories across Canada. Informants included individuals with ACHD and families ( $n = 7$ , ages 24-72), and practitioners located in disciplines including cardiology ( $n = 6$ ), nursing ( $n = 3$ ), cardiovascular surgery ( $n = 1$ ), psychology ( $n = 2$ ), dietetics ( $n = 1$ ), and kinesiology ( $n = 1$ ). Half of informants were involved in research and almost all of them identified themselves as advocates. The practitioners ranged in

**TABLE 1** Key Canadian contributions

Estimated dates	Contribution
Early 1800s	Paper by Dr. Andrew Holmes was referred to as one of the first publications describing an adult with complex CHD who died at 23
Late 1800s-1936	Dr. Maude Abbott (curator in the pathology building at McGill University) examined the heart noted above and hundreds of other specimens with heart malformations. She helped classify CHD and her work was published in Osler's "modern medicine textbook" and then in 1936, she "published the Atlas of Congenital Cardiac Disease"
1959 and onward	The Adult Congenital Cardiac Clinic was founded in 1959 and formalized in 1982 at Toronto General Hospital. It was the first, and remains the largest clinic in the world for ACHD care and is known as a template for other organizations and a hub for patient care, education, and research
1990	A turning point was the meeting in Bethesda, Maryland in which practitioners and researchers assembled to discuss the problem and lay out strategic steps for ACHD
1991 and onward	Canada is known as a leader in ACHD care and training of fellows. The Canadian Adult Congenital Heart Network formed in 1991 which now has 15 congenital clinics and 8 cardiac congenital, surgical programs. Agreement that these regional centers of excellence are the best model of care and acted to improve care. The ACHD Learning Center formed with Canadian representation on the board and the addition of the iHeart Change website
1999-2010	Continual shift from trial and error to management approaches informed by evidence. Canada has been a leader in the management of ACHD with the first worldwide guidelines in 1996 and more recent guidelines in 2009 and 2010

experience with the congenital population from 2 to 50 years. Ethics approval was received for the study.

### 3 | FINDINGS

We present the findings according to 6 main areas related to treatment: (1) Prior to 1990; (2) The Organizational Beginning; (3) 1990 and Onward; (4) Transfer to Adult Care; (5) Comprehensive Approaches; and (6) Demographic Shift. See Table 1 highlighting key Canadian contributions and Table 2 highlighting summary of future directions. In the findings, we differentiate quotes for health care professionals with (hcp) and patients/families with (pt/f).

#### 3.1 | Prior to 1990

Historical foundations of ACHD treatment date back to the 1800s. One informant noted, “correlation of anatomical findings of CHD goes back to the early 1800s” (hcp). A paper by Dr. Andrew Holmes was referred to as one of the first publications describing “an adult who died at 23 ... found [to have] a complex CHD” (hcp). Almost a century later, this heart was examined by Dr. Maude Abbott at McGill University, who was “a curator in the pathology building” (hcp). Under Sir William Osler’s encouragement, she examined “hundreds of specimens” with “heart malformations” (hcp). Her work, published first in Osler’s “modern medicine textbook” and then in 1936, she “published the Atlas [of Congenital Cardiac Disease],” which helped “classify CHD, the nomenclature” (hcp). Her work paved the way for innovative treatments that began in the 1940s with children and then, more so in the late 1950s and 1960s with adults when the “capacity to do open-heart” (hcp) surgery on cardiopulmonary bypass had developed.

Treatment was described as “haphazard until 1990” in which practitioners “were pioneers” using “trial and error” because there was no empirical evidence, and oftentimes the procedures had never been done (hcp). One informant described, “going by the seat of your pants because we didn’t have a long record of treating adults.” Another informant recalled incising the chest and opening the rib cage noting, “I had no idea what I was looking at so I asked my colleague, and he had no idea either ... that was

what was happening” in the early days of treatment. Practitioners relied on previous experiences and collaborative discussions to guide clinical decision making, “we met weekly on how to manage these patients ... in many ways the decisions were made by group intuition.” Another informant stated, “in a lot of cases” treatment and surgical effectiveness is still not known, “but we know more than we used to” (hcp). Treatment evolution was complicated because you could not “make broad generalizations ... [it is] challenging, no two patients present the same, no two diagnoses are the same” (hcp).

Patients and “their families, consented” to these unproven procedures and “a lot of lives were lost as these surgeries were being developed and evaluated” (hcp). Referring to her child who was born with CHD 30 years ago, 1 informant referred to “never ... [being] given a lot of hope.” The informant reflected on what practitioners relayed to her:

We can repair it ... it might give two years to her life. ... and then when the Fontan was done, well she might live to age 10 but nothing exists after that. ... It was like that, all the way, so I couldn’t even think in my head of, *adult congenital*, because it sort of did not exist.

More recently, an informant referred to a surgery completed on her adult child. She was told, “[the procedure] is not done on congenital, until very recently ... then you’d sort of Google how many were done, how many survived? I thought ‘oh God’ it’s really just experimental.”

#### 3.2 | The organizational beginning

A turning point was a “meeting in Bethesda, Maryland in 1990” (hcp) organized by the American College of Cardiology. This meeting “marked the organizational beginning of ACHD” and was “the first time that experts got together to define the problem” (hcp). In 1991, the “Canadian Adult Congenital Heart Network” was formed which has “15 congenital clinics ... 8 cardiac congenital, surgical programmes” (hcp). Following that, The International Society for ACHD was founded, which has had 4 Canadian Presidents. Canada has been at “the forefront ... Toronto, Montreal, Vancouver and other places have been part of all the important organizations” (hcp). Many

**TABLE 2** Summary of future directions

Future directions	Challenges and strategies
Evidence-informed approaches	The shift to evidence-informed approaches has been complicated by the complexity and heterogeneity of ACHD. Research and expertise coupled with the computerized database has led to an emerging body of evidence and multiple guidelines
Philosophy of patient-centered care	The engagement of patients as active partners in their care has been aligned with the general shift in the health care system. This philosophical approach can be strengthened by the continued modification of practitioners’ perspectives and dedicating more time to engaging patients
Comprehensive approaches	Additional resources are required to optimize the comprehensive approach to care so that it includes sufficient access to services such as psychological support, cardiac rehabilitation, dental care, and support networks
Gap in resources	There is a significant gap in resources accompanying the demographic shift. Advocacy and government leadership are required in order to recognize ACHD as a lifelong illness and secure sufficient funding

informants referred to Toronto's clinic as "one of the oldest and largest programmes in the world," a "template for other organizations," and "a hub for patient care, education, research" (hcp).

Adult congenital heart disease emerged as a "sub-specialty of cardiology" (hcp). Informants explained, CHD is "a specialist disease" requiring "experts" who understand the "special anatomy" because "everything is different, the heart connections are completely abnormal. And then, the surgeons modify the anatomy again" (hcp). Adult congenital heart disease was described as having "a lot of subtleties to the management ... requiring a great deal of experience and a great deal of knowledge" (hcp). There was general agreement among health care professionals that "regional centres of excellence" were the best of model of care and helped "concentrate the experience in a limited number of people and to have those people collaborating." This concentration of experience within the "network of adult congenital heart clinics" is the "basis of improved care" and one reason that Canada is known as a "leader" in ACHD care (hcp). One health care professional referred to a paper that provided evidence "that ACHD patient care in special facilities reduces mortality and improves outcomes."

Problematic is that "75%" of people with ACHD are not being seen at "specialized centres"; "they are either not followed or they are followed by general practitioners" which is "not fair to the patients, it's not fair to the general practitioners to have to look after some weird and wonderful congenital heart patients" (hcp). As a result, these patients "are referred with major complications which are difficult to manage" (hcp). Although informants were explicit about "receiving care from someone with specialist ACHD training," they also noted, "there are not enough of those people to go around" (hcp). Although there was some variability, 1 informant specified, "wait-time for a new patient at [their] clinic is two years" (hcp). Thus, some clinics focus on "complex CHD" and have those with "simple lesions ... seen by community cardiologists" (hcp). Informants noted an important need to "liaise with community partners" including "family doctors, the primary caregivers and the community cardiologist" (hcp). Initiatives such as "education days for community providers" as well as the "ACHD Learning Center" helped "bridge the knowledge gap in the community" (hcp). Building these collaborations was helpful "so that patients could see a local community cardiologist where they live" (hcp).

On-the-job learning was typical until "speciality training" for cardiologists began in "1990" and is now, offered in "Toronto ... Montreal, Edmonton and Vancouver" (hcp). It was noted, "Canadian training programmes have strengthened over the last decade" and are involved in training "ACHD specialists from around the world" (hcp). A concern voiced by patients was the belief that fellows "coming through are taught today's treatments ... aren't well educated in procedures that they don't do anymore." One informant commented, "every time I go to the hospital I have to tell someone how to take my blood pressure ... you can't take blood pressure in either arm, I have shunts." Patients often referred to themselves as "walking encyclopaedias of what the advances in cardiac care have been." It was noted that "on-the-job training" (hcp) remains typical

for practitioners located in disciplines such as nursing, psychology, dietetics, and kinesiology. One individual noted, it is mainly "self-directed learning ... you have to build your own capacity" about CHD. Informants described "seeking out different conferences," and "mentorship and conversations with other practitioners."

### 3.3 | 1990 onward

There has been a shift to "evidence" informed management approaches in which Canada has been a leader in ACHD management with the original 1998 Canadian Cardiovascular Society Consensus Conference report as well as more recent guidelines (2009 and 2010) published in the Canadian Journal of Cardiology. One informant indicated, "Canadians have been part of all the important guidelines" (hcp) with representation on the European and American guidelines and those specific to pregnancy and ACHD. It was noted, the "new American guidelines" for ACHD management will be "focus[ed] on evidence" (hcp) One informant remarked that the focus of the first guidelines was, "this is what the experts in the field do—because there was no evidence, so all you had was peoples' experiences" (hcp). However, it was noted, there is still "no evidence" for "many things" ... but experience is very important" (hcp) in clinical decision making.

Data and evidence were identified as vital to the expanding ACHD knowledge base. One initiative was "a computerized database" and formal tracking system called the "Congenital Heart Surgeons' Society Data Center" (hcp). This center pools data and tracks population outcomes across "75 institutions in North America" and offers "sophisticated statistical analysis ... [of] a critical mass of patients" (hcp). A noted shift was "measuring survival to measuring morbidity and more recently, functionality" (hcp). Another informant referred to "a coding system for diagnosis" so that "sites used the same diagnostic codes" since around 1990, which provides opportunities to "collaborate on research and get statistics." (hcp). Informants remarked, "multi-site research is critical in order to ... track outcomes so that treatments can be evaluated" (hcp). It was also noted that "randomized controlled trials [are] not always realistic" because this cohort "is very heterogeneous" and thus, there are not "thousands of patients with a specific diagnosis to do an RCT ... you end up with no numbers. Then, you don't have the power to make conclusions" (hcp). Additionally, the ACHD cohort is often "excluded" because they can "skew the [cardiac research] data" (pt/f). As the system moves forward, informants noted, "[it is important] to figure out how we best handle the really complex patients ... what innovations we can pursue," exploring "outcomes of people who are and are not in specialized ACHD care," and "asking the patients themselves what they'd like to see" (hcp).

Treatment has advanced substantially in terms of technology. One informant commented, ACHD "is where pediatric CHD was 30 years ago. ... We're kind of just on the precipice" (hcp). Informants referred to refinements in surgical procedures, "advancements in medical technology and diagnostic imaging," "three-dimensional modelling," "interventional cardiology," and "telehealth" (hcp). Although there

were issues associated with “privacy and liability” across provinces, this last item was identified as important for enhanced accessibility with “remote” settings (hcp). It was noted that the above areas will continue to advance over the next 10 years as well as advancements in a range of other areas such as “personalized medicine,” “bioengineering,” “electrophysiology,” “neuropsychology testing,” “heart failure treatments,” and “less invasive procedures” (hcp).

Informants remarked on the importance and “expansion” (pt/f) of interdisciplinary teams (eg, nurses, nurse practitioners, cardiologists, surgeons, interventionists). It was noted that “the multidisciplinary aspect ... was a centre piece” of 1 clinic’s plan because “many players” are required to “provide care well” (hcp). Considering the unique attributes of this cohort, informants described the significance of “group-think” in which “different disciplines bring a different knowledge to the table” (hcp). Some of the practitioners specialized in “reproductive health,” “electrophysiology,” “genetics,” “heart failure,” and “palliative care” (hcp); however, access to psychiatrists, psychologists, and social workers was severely limited and variable across Canada.

### 3.4 | Transfer to adult care

Transfer to adult care at 18 is typical because CHD requires “long-term care” (hcp). However, 1 informant indicated, “until [the] late 1990s, early 2000s, there was no organized transfer” (hcp) to adult programs and many individuals with CHD were “lost to follow-up.” Prior to formalization of transfer programs, 1 informant described feeling, “not supported, you’re like ‘Oh my god’ I don’t know anyone” (pt/f). Informants indicated that transfer could be better formalized so that it gave “you a sense of power and control over your healthcare” (pt/f) and “a renewal of understanding ... so that they may feel more independent and supported” (hcp). Another informant (family member) indicated, “the transition was difficult ... but transitions are always difficult ... not knowing anyone I could call.”

Informants remarked on the major enhancements of transfer programs: “there’s been leaps and bounds in transition” and now described as “simple” (pt/f). One informant explained that transfer to adult care in their program is “done with the pediatrician available for that first interaction ... because I am a stranger” (hcp). Another informant described “a transitional evening” where “a multidisciplinary group ... build relationships ... [with] parents and patients” (hcp). However, this evening is offered a limited number of times per year and some people have difficulty traveling to the hospital. Many informants referred to the positive role of nurses during transfer, stating: “my congenital nurse was amazing, once I was under her care ... it was a world of difference.” Another informant referred to “the iHeart Change website” as “a great resource ... to direct patients” to for “questions” and “connections in the community” (hcp).

Although less of an issue, there are still “patients lost to follow-up” (hcp). One informant remarked, “perhaps pediatric patients ... feel, you’ve had your procedure, you’re fixed” (hcp). It was noted, “more awareness” is needed to reinforce that CHD “is a lifelong concern ... women might be pursuing pregnancy, or acquired heart

disease risks as life goes on, and how will those affect you” (hcp). The culture of adult care is different in comparison to pediatric care: “there’s not much interaction with other patients in the waiting room ... you get to adult care and it’s like number 84” (pt/f). Another young adult stated, “I have to like stand my ground that I want my mom around.” The informants also noted there is a sense that “my word is good now ... because I’m an adult” (pt/f).

### 3.5 | Comprehensive approaches

Unique features of ACHD treatment are the need for “holistic” (pt/f) and “comprehensive approaches” (hcp). Although ACHD care is “highly specialized ... [it is a] multi-system disorder” (hcp). Practitioners also assess and discuss: “psychosocial issues,” “obesity,” “physical activity,” “healthy diet,” “complications,” and “reproductive issues” (hcp). Another informant remarked, “[it is] a dangerous medicine ... to just focus on a small area, and don’t see that this patient actually does have other problems” (hcp). ACHD “affects your entire life” from “career choices to family to where you live” (pt/f). There has been recent emphasis placed on “quality of life” (hcp). Informants indicated that care will be even “more holistic” and focused on “lifestyle” in the future as well as research initiatives that look closely at “what is their quality of life?” and “are there things we can do to improve their quality of life?” (hcp).

Psychological care was a significant gap in the provision of care with a substantial lack of access to social, psychological, and psychiatric care. Informants commonly stated, “we don’t have a counsellor ... a psychologist ... a social worker” (hcp). Psychological care was “missing from most places” and rarely was it “fully integrated” into clinics (hcp). This was problematic considering “there’s a big psychological component” to CHD and issues such as “depression” and “coping with a new diagnosis, or adjusting to declined health status, or end-of-life concerns” (hcp). This population “are at higher risk of anxiety and mood struggles ... on top of regular life stressors, so family, finances, relationships, this is another challenge that gets added on top” (hcp). One informant explained when undergoing a procedure, there are many “extra cares” to think about: “your spouse, your mortgage, you’ve got all these extra cares, how long am I going to be gone? how long is the rehab? I can’t get life insurance. Can I return to my same job? Do I have the emotional, do I have the physical, do I have the stamina to return to my work?” (pt/f). One informant noted “when we talk about medical outcomes, the people who are not depressed and not anxious tend to do better” and yet, “mental healthcare is not a priority at the government level” (hcp). Many provincial and territorial insurance plans do not reimburse for these services. In moving forth, a “major investment at the leadership, and that’s including the provincial level ... the healthcare system” (hcp) is needed.

The “philosophy of patient-centred care” (hcp) has been identified as important. Informants commented, “we look at their social situation, their mental health status ... their individual characteristics” (hcp). On a whole, patient/family informants generally found practitioners to be “compassionate ... empathetic.” One informant stated, “I would tell her about a silly problem and she would actually call me

back and discuss it." It appeared that historically, practitioners were less concerned with patient engagement. One informant stated, "I got a tubal ligation ... at the time, it was just 'this is what you have to do' and I was like 'okay.' Like it wasn't really a decision." Informants generally indicated a "big difference throughout the years" in terms of "engagement," "empowerment," and "having patients involved in decision making" about "their management plan" (hcp). One informant commented, "conversations are more open ... where they share their knowledge and I give them feedback from my experience of my own body" (pt/f). Another informant remarked that practitioners will "listen to me ... I feel like I have more control, more power" (pt/f).

In moving forth, informants identified other areas that required attention to further optimize comprehensive approaches. For example, there was a "lack of dental care" (hcp) and access to cardiac rehabilitation (CR). It was noted, there is a "waitlist" for CR as well as "resistance" (hcp) from CR settings when the patient is not surgical or had not had a myocardial infarction. Sometimes the resistance is related to "a level of comfort" because there is "a lack of awareness" about CHD and there are no "best practice guidelines" on how to "prescribe exercise" in this population (hcp). Although informants noted being "more actively aware of the need for palliative care and management earlier ... there is still resistance" noting that "we don't do it very well" (hcp). The informant explained, "it's very hard to switch gears from, things are going to get better to things are not going to get better" (hcp). Another gap was the "lack of support networks" (pt/f) and the need for "peer support" (hcp). Oftentimes, this cohort "feels like freaks. They're isolated. And to have a community is important" (hcp).

### 3.6 | Demographic shift

Informants repeatedly used words such as "tsunami" to refer to the ACHD patients who are "surfacing ... in large numbers" (hcp). It was noted that ACHD used to be "rare" because this cohort "never existed" (hcp); mortality occurred in utero or as newborns. "Sixty years ago only about twenty per cent of patients ... survived into adulthood" (hcp). These statistics have dramatically reversed with "ninety per cent of these patients surviving" (hcp). One informant said, "for the first time in history, there are more adults than children" living with CHD (hcp & pt/f). This demographic shift involves a "constant influx ... the numbers are going to continue to rise" because "the influx is greater than the death rate" (hcp). It is noted that "one out of 100 babies born has a congenital heart defect" making it "the most common congenital defect" (hcp).

It was articulated that the unique and complex nature of CHD requires life-long, specialized care: "The hearts of these patients are not cured ... they have long-term issues" (hcp). For example, a mild condition may require "monitoring ... every two to five years ... non-invasive investigations such as an ECG and an ultrasound of their heart" (hcp). The severe end of the spectrum is a child born critically ill with multiple "open-heart operations" and "interventions" and transfer into adult care with "a large burden of chronic illness" requiring "frequent surveillance" (hcp). Complicating the matter

is that these adults "might have issues related to neurocognitive development ... trouble completing school ... have not had stable families ... difficulties even with drug abuse" (hcp). An example of a complex case may be an adult with "mechanical heart valves" who "needs to take blood thinners every day, go for blood tests every month. They can get infections on their valves, they can get clots on their valves" (hcp). As 1 informant explained, these patients can "slowly deteriorate and eventually need a heart transplant" (hcp).

The significant shift in the demographic profile of the person with CHD is remarkable, but it "is hitting the [adult] healthcare system" (hcp). There is "a huge gap" because outpatient volume and inpatient admissions "have increased exponentially without of course funding [the] increase in resources" (hcp). Key informants across Canada resoundingly noted that "it's a system that's inadequate for the need," the clinics are "under-resourced" and practitioners are "overwhelmed" and "working beyond capacity" (hcp). Another informant noted, "patients are knocking at our door, they have problems, and then, everybody is stretched to the limits" (hcp). Informants remarked, "more providers" and "more support for providers" is required to address the demographic shift (hcp). Complicating the matter is ACHD disease complexity. One informant commented seeing "only one patient with CHD" in the time that "three patients with acquired heart disease" (hcp) can be seen. A first clinic visit can involve reviewing "hundreds of pages of medical history ... to understand the full history, and it can be challenging to come up with the right treatment plan" (hcp). Informants remarked that these patients are "time and resource consuming," "an expensive oddity ... a cost ... surgeries take longer," and "the healthcare system ought to recognize that" (hcp).

The complexities embedded in this demographic shift are influenced by not being able to predict the future in terms of a disease that was generally limited to children. One informant noted having limited resources "because who could develop the resources before knowing what really the future was going to hold?" (hcp). This informant elaborated, "the future is still happening ... [however] there's a huge problem now where there's not enough resources and it's not being funded by the Ministry. ... the question [now] is how to get the message to the decision makers." It was apparent that despite the demographic shift, the resources and funding for the adult population are not equivalent to pediatrics. Complicating this matter is the "ethical issue" (hcp) of dwindling resources for adults. One informant stated that treatment is provided to "an infant that was destined to not make it, and out of that creates a 29-year-old woman who is married, with a husband, with a job, has a university education and she's having kids" (hcp). The predicament is described as:

[Now] she needs another operation ... can we say "oh, well" we gave you 29 years, just get sick and die, leave your kids as orphans when you're 35. ... the question is, is it society's responsibility? And society gets translated into our government and into the Ministry of Health ... And I'm going to say "yes" because if you're going to be true to the decisions that were made



when you said “yes, we’re going to treat these babies,” it’s the right thing to do.

This ethical quandary challenges society’s values as well as equitable funding distribution to account for the drastic demographic shift. An informant said, “We are the problem, we are the adults with this disease ... we’re grateful this problem is there” (pt/f). Informants highlighted the need for “advocacy” (hcp & pt/f) and “government leadership”—we need to “educate the leaders, the Minister of Health, the politicians about this increasing number ... the complexity and long-term problems” (hcp) of this cohort.

## 4 | DISCUSSION

Our research reflected the pioneering efforts of Maude Abbott who had a substantial impact on the evolution of ACHD. Evans and Béland<sup>12</sup> identified Abbott as laying the foundation for cardiac malformations in the early 1900s when she examined and catalogued hundreds of pathological specimens. Although this historical investigation has demonstrated an overall shift to evidence-informed treatment based on an emerging body of research, the nature of this shift remains complicated by ACHD’s complexity and heterogeneity. There is some resistance to traditional intervention research because of this heterogeneity as well as the need for group intuition and experience to guide clinical decision making with this cohort; we do however, recognize the latter elements as one form of evidence. Our work also echoes what others are saying in terms of the need for both multi-site trials and large data sets that can be facilitated through data registries.<sup>3,13</sup>

The emerging body of evidence has led to multiple guidelines concerning ACHD management.<sup>14</sup> Our work supports others who have indicated that specialized care of this population is fundamental<sup>15</sup> combined with care from community cardiologists and primary care providers. The need for specialized care of the ACHD population was recognized in the 1980s<sup>3</sup> and has since been found to reduce mortality.<sup>15</sup> We have extended this work by highlighting how specialized care was advanced with the organizational beginning of the 1990s, which also promoted collaboration of central bodies and formalized ACHD practitioner training.

Problematic is that many adults are not routinely followed by practitioners specializing in CHD<sup>16</sup> and/or by 1 of the 15 ACHD clinics in Canada.<sup>17</sup> There simply remains inadequate numbers of trained specialists<sup>18</sup> despite the mounting cohort of adults with CHD. A Pan-Canadian study reports that national wait-time targets are not being met.<sup>17</sup> This issue is complicated by the loss to follow-up that occurs when children are not transferred to adult providers<sup>19</sup> and comprehensive, formalized programs are not in place to ensure smooth transfers from pediatric to adult care.<sup>20</sup> Our work, among others,<sup>21</sup> have consistently indicated that this is problematic because the unique, complex and multisystem nature of ACHD requires multidisciplinary and specialty-trained teams. Additionally, there is insufficient access to psychological and social support provided by social workers and psychologists<sup>17</sup> as well as a need to integrate

psychology into ACHD programs.<sup>22</sup> Our study underlined the significance of this issue both from the patient and provider perspective who emphasized the social and psychological impact of ACHD that interferes with daily functioning.

Invariably, an issue that we grappled with is the misalignment of resources. It was clear in this historical investigation that the number of adults requiring specialized services continues to rise while resources continue to lag. Similar to others,<sup>16</sup> our research highlighted the significant gap in available inpatient and outpatient resources to treat the rising ACHD population. Our work echoes others about the need for lengthy assessments including reviewing clinical notes that often date back to birth because current issues with adults are related to previous interventions.<sup>21</sup> It is expected that costs will continue to rise considering that the ACHD population is only getting bigger. Over a 10-year period, it was found that there were increased number of hospitalizations for adults compared with children<sup>23</sup> and that over time, the increase in costs were higher for adults compared with children who have CHD.<sup>24</sup> Our work echoes the urgency for politicians in the health care arena to recognize the scope of this problem so that adequate funding is achieved.<sup>1</sup>

## 5 | CONCLUSION

There are key Canadian contributions to the advancement of ACHD care that have extended the life expectancy and enhanced quality of care for this population. Political leadership and financial commitment are paramount to maintain Canada’s role as a global leader in caring for the mounting population of adults with CHD. Researchers have highlighted a significant inadequacy of resources for the number of adults living with CHD, and as a result, the real risk of premature death for the ACHD population.<sup>1</sup> Health care system changes are necessary so that resource allocation is prioritized in this population. These insights behoove the community of stakeholders to think creatively on how to draw attention to this issue and the unique and diverse needs of this population.

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## CONFLICT OF INTEREST

None.

## AUTHOR CONTRIBUTIONS

*Concept/design:* JL, SF

*Recruitment and data collection:* JL, SF, BB, TY, JG, AM

*Drafting of article:* JL

*Data analysis/interpretation, critical revision, and approval of the article:* JL, SF, BB, TY, JG, AM.

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