Adult Congenital Heart Disease—Preparing for the Changing Work Force Demand

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KEYWORDS

• Congenital heart disease • Health care • Life course • Noncardiac surgery

KEY POINTS

- The demographics of the ACHD population continue to evolve and require a life course approach to policy and workforce planning.
- Adult congenital heart disease is a multisystem condition encompassing not only the congenital heart defect but the additional cardiac and non-cardiac morbidities that may interact with the underlying CHD.
- Establishing methods to provide, measure and improve high –quality healthcare is imperative to maintaining patient care and the appropriate workforce for adults with CHD.

INTRODUCTION

This volume is dedicated to advances in the care of adults with congenital heart disease. Scientific inquiry is new to our field relative to other forms of cardiovascular disease in adults. Non-the-less the last 2 decades have seen an exponential increase in the number of guidelines generated in several international expert groups, most recently in the US population data from a variety of jurisdictions and single center studies demonstrating the science behind the success of our field.[1] As we enter this third decade of the twenty-first century, there is a need to assemble the emerging data on several specific conditions that mandate the attention that longevity now permits. In this chapter the authors review the data cornerstone to the growing workforce needs. They first review the determinants of disease in congenital heart disease (CHD), outlining the demographic changes that underlie the growing population in terms of number of children compared with adults, the age distribution of the population, and the emerging sex differences. They propose a life-course epidemiology framework to capture the complexity of a condition expressed variably over the stages of life. The authors then summarize the rapidly growing body of evidence that CHD in adults is a multisystemic condition with rising complication rates as patients age and develop complications at a distance from the heart. They then review quality indicators specifically developed for adults with CHD as applied to recommendations for follow-up at the patient level. This first chapter serves as a backdrop to the Susan M. Fernandes and colleagues' article, "Access and Delivery of Adult Congenital Heart Disease Care in the US: Quality Driven Team Based Care," in this issue that applies these observations to the planning of health care services delivery in the United States accounting for the definition and organization of multisystem expertise and centers for adults with CHD at a health systems level.

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Cardiol Clin 38 (2020) 283–294
https://doi.org/10.1016/j.ccl.2020.04.011

DETERMINANTS OF HEALTH AND DISEASE IN CONGENITAL HEART DISEASE—WHAT ARE THEY?

Demographic Shifts in Age, Disease Severity, and Sex

The changing age distribution of CHD provides a powerful motivation to reorient the workforce, as demographic shifts are resulting in a growing number of adults and women with CHD. Data supporting the shift in demographics began to emerge between 2000 and 2010. On a population level, a rising prevalence of CHD in adults compared with children from 1985 to 2000 was observed, such that the number of adults and children with CHD had equalized as shown in Québec and expected to reflect national and international trends.[2] By 2010, the number of adults with severe as well as all forms of CHD exceeded the number of children, with fully two-thirds of the population being adults.[3] Using comprehensive population data sources, the life span prevalence rates of subjects with CHD were documented: 8.12 per 1000 at birth, 13.11 per 1000 in children, 6.12 per 1000 in adults, and 3.7 per 1000 in geriatric populations older than 65 years, thus warranting coining the term "Geriatric Adult Congenital Heart Disease."[4]

In the United States, working together with the Center for Disease Control and Prevention (CDC), investigators used Canadian data to estimate the magnitude of the problem, generating first-time empirical estimates. **Fig. 1** illustrates that of a total of 2.4 million subjects in the United States with CHD, 1.4 million were adults, whereas 1 million were children in 2010.[5] **Table 1** illustrates the age distribution of CHD in the United States with prevalence rates and corresponding numbers by severity of disease. Obtaining prevalence rates of 6.16 per 1000 adults older than 18 years, there were, as generated by empirical estimates in 2010, a total of 160,000 adults with severe CHD compared with 123,000 children with severe CHD underscoring the need to expand the workforce needed to care for adults with both simple and complex forms of CHD. In the United States, several methods have been and are being used to generate direct empirical measures of adult CHD (ACHD) populations.[6,7] Using capture and recapture methodology applied to 3 inpatient and outpatient data sources in 11 New York counties from 2008 to 2010, the prevalence of adolescents with CHD was 6.4 per 1000 cases.[8] Using similar methods applied to administrative records in Atlanta, Georgia counties, a prevalence rate of 6.08 per 1000 adults older than 20 years was observed in 2010.[8] Using ICD-9 codes and data sources in Atlanta, Massachusetts and New York, the proportion of patients with severe CHD seeking health care varied between 13% and 21%, which is higher than previously observed more conservative estimate of an approximate 10%.[5,7]

An important observation in population-based surveillance studies is that despite the limitations in ascertainment of gender-related health care behavior, there seems to be a predominance of women among adults with CHD, notably in the reproductive age group. **Fig. 2** illustrates a striking predominance of women compared with men in the 18- to 45-year-old age group. In 2010, there were 342,000 women compared with 236,000 men with CHD in the United States, with a higher proportion of women with severe CHD than men.[5] As part of the larger CDC surveillance initiative, 5672 pregnancies were identified in 26,655 women with CHD between the ages of 11 and 50 years. Interestingly, age-adjusted pregnancy rates did not substantially differ between women with severe and nonsevere CHD and were observed to be between 10% and 25%.[9] Moreover, pregnant women with CHD of any severity had more noncardiovascular comorbidities.[9] Sex differences in outcomes and health services utilization have been shown between men and women using the Kids Inpatients Database in the United States in children and using the Dutch CONCOR database and the Québec CHD database in adults. As where more infant men undergo high-risk cardiac procedures, surviving women have less severe forms of CHD, potentially optimizing reproductive fitness.[10] The protective effect conferred to adult women carries over into adulthood in terms of health services utilization underscoring the importance of gender-driven variations in health behavior that can affect cardiovascular disease outcomes.[11,12] Thus, there is ample empirical evidence to support the lifespan demographic framework of CHD where the same lesion expresses it differently depending on the life stage. This is so not only from childhood to adulthood but also across the developmental frames that adulthood now spans: from the young adult, to the adult of reproductive age, to middle age, and finally to advanced age.

A Life Course Epidemiology Framework

Thus, a conceptual shift is needed to consider the implications of demographic shifts in CHD across the lifespan. This poses a particular challenge on the growing workforce requirement where interdisciplinary care models need to be integrated not just vertically from primary care to specialized

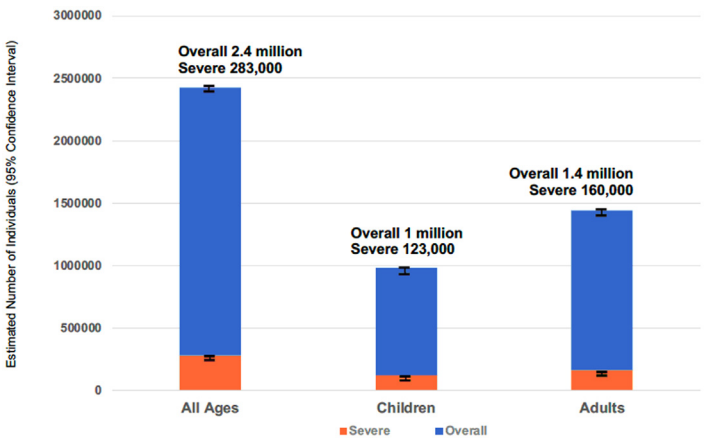

Fig. 1. The total population of CHD in the United States shown by age in children compared adults for all and for severe CHD in 2010. Corresponding prevalence rates per 1000 (CI) for all ages-overall 7.85(7.79–7.92), for all ages-severe 0.92(0.90–0.94), for children-overall 13.21(13.03–13.39), for children-severe 1.66(1.60–1.73), for adults-overall 6.16(6.10–6.22), and for adults-severe 0.68(0.66–0.70). (*Data from* Gilboa SM, Devine OJ, Kucik JE, et al. Congenital Heart Defects in the United States: Estimating the Magnitude of the Affected Population in 2010. Circulation. 2016;134:101-9.)

care but horizontally as persons with CHD progress from one life stage to another. Fig. 3 outlines a life course epidemiology framework illustrating the developmental time frames for the expression of CHD across the lifespan.[13] Life-course epidemiology moves beyond longitudinal studies, bringing theoretic constructs that facilitate the modeling of disease events across the lifespan, improving study design and analyses.[13–15] The life course health development framework was created to shift the emphasis away from disease and toward health, with the knowledge that health is a consequence of genetic, biological, and social determinants and with the understanding that health development is an adaptive process.[14] As illustrated in Fig. 3, as patients with CHD move across the life stages, determinants of health include genetics, CHD severity, psychosocial health, health behavior, and the environment in addition to access to the right health care. These converge with processes of health development to determine developmental trajectories of health and expression of disease across the lifespan.[13]

Table 1
Estimated prevalence and numbers of congenital heart diseases by age, severity, and race-ethnicity in the United States in 2010

Category and Age Group	CHD Severity/ Race-Ethnicity	Estimated US Prevalence per 1000 (95% Confidence Interval) (%)	Estimated No. of Individuals (95% Confidence Interval)
CHD severity			
All ages	Overall	7.85 (7.79–7.92)	2,425,000 (2,405,000–2444 000)
	Severe	0.92 (0.90–0.94)	283000(277000–290000)
Children	Overall	13.21 (13.03–13.39)	980000 (966000–993000)
	Severe	1.66 (1.60–1.73)	123000 (119000–128000)
Adults	Overall	6.16(6.10–6.22)	1444,500 (1431,000–1459,000)
	Severe	0.68 (0.66–0.70)	160000 (155000–165000)
Race-ethnicity			
Children	Non-Hispanic white	13.31 (13.12–13.49)	620000(612000–629000)
	Non-Hispanic black	12.69 (12.50–12.88)	133000 (131,000–135000)
	Hispanic	13.26 (13.08–13.45)	227000(224000–230000)
Adults	Non-Hispanic white	6.36 (6.29–6.42)	1,104,000(1 094000–1115000)
	Non-Hispanic black	5.63 (5.56–5.69)	155000 (153000–156000)
	Hispanic	5.58(5.52–5.65)	186000 (184000–188000)

Children are those aged 0 to 17 years and adults older than or equal to 18 years.

From Gilboa SM, Devine OJ, Kucik JE, Oster ME, Riehle-Colarusso T, Nembhard WN, Xu P, Correa A, Jenkins K and Marelli AJ. Congenital Heart Defects in the United States: Estimating the Magnitude of the Affected Population in 2010. Circulation. 2016;134:101-9.

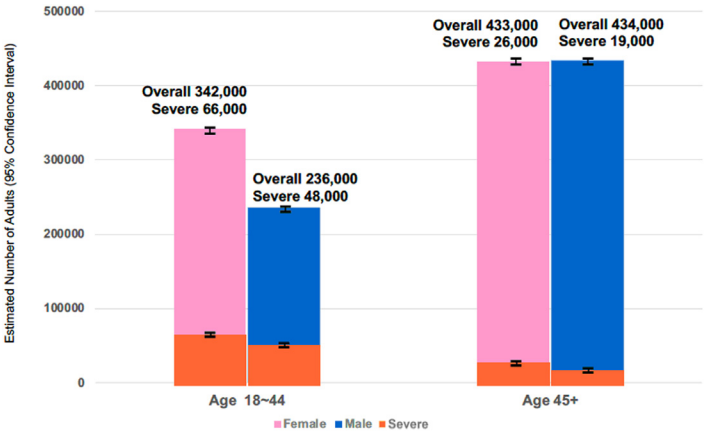

Fig. 2. The distribution of CHDs by sex showing a predominance of women of reproductive age compared with men in the United States in 2010. Corresponding prevalence rates per 1000 (CI). Women:18～44 overall: 6.10(5.92–6.29), 18～44 severe 1.19(1.10–1.27), 45+ overall 6.70(6.58–6.81), 45+ severe 0.41(0.38–0.44). Men: 18～44 overall: 4.24 (4.08–4.40), 18～44 severe 0.88(0.81–0.96), 45+ overall: 7.59(7.46–7.72), 45+ severe: 0.33(0.30–0.36). (*Data from* Gilboa SM, Devine OJ, Kucik JE, et al. Congenital Heart Defects in the United States: Estimating the Magnitude of the Affected Population in 2010. Circulation. 2016;134:101-9.)

Shifting Mortality—Can We Do Better?

Advances in surgical and clinical management of CHD have allowed more than 90% of children to survive to adulthood, but this development has led not only to a shift but also to a swelling in mortality into adulthood.[16] Consistent with these findings, in Belgium, an analysis of survival trends by cohort and defect type was performed using administrative and clinical records. This showed that overall survival to 18 years of age for children born between 1990 and 1992 was nearly 90%,

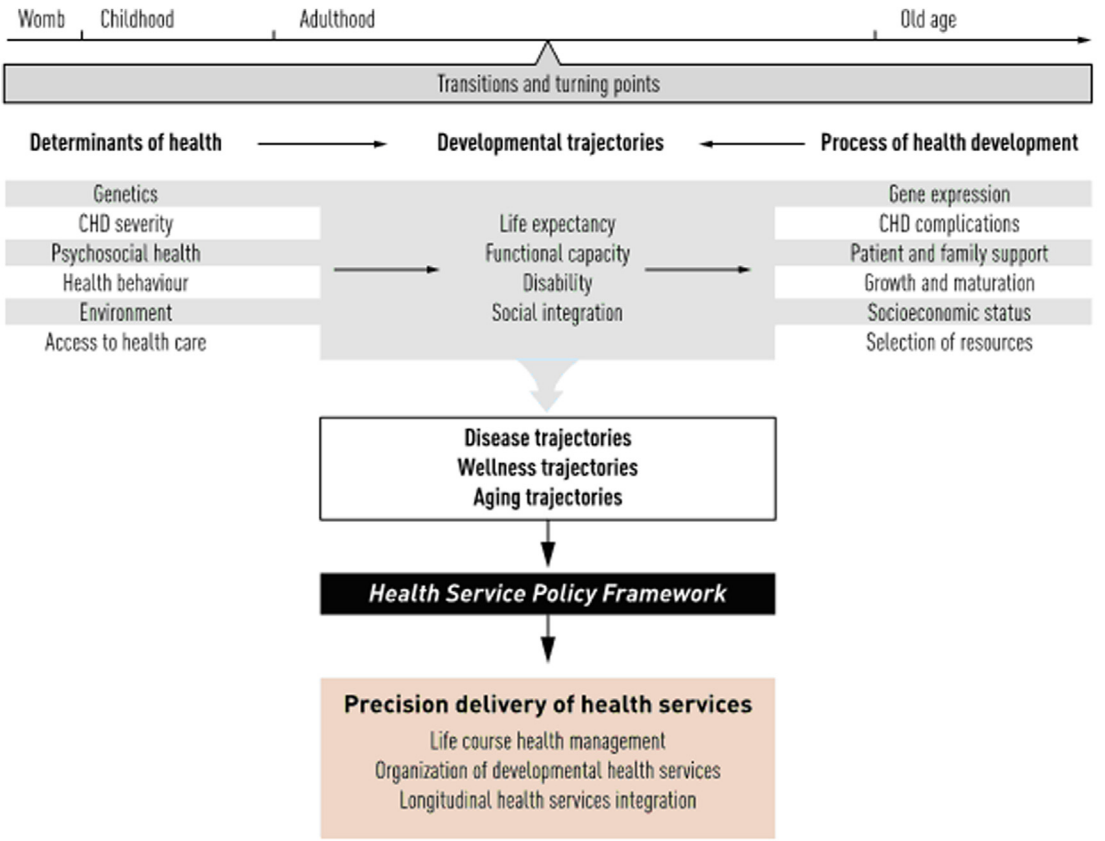

Fig. 3. Life course epidemiology framework illustrating the developmental time frames for CHD. (*From* Marellli AJ. Trajectories of Care in Congenital Heart Disease- The Long Arm of Disease in the Womb. Journal of Internal Medicine. 2020; in press.)

showing significant improvement compared with previous decades.[17] Notwithstanding this progress the question remains: is the glass half empty or half full, and what can we do about it?[18] Despite major progress in CHD mortality due to surgical interventions in children and demonstration that specialized ACHD care can improve mortality where universal health insurance optimizes access,[19] cohorts of patients with ACHD did not show the same degree of benefit as children with CHD between 1973 and 1993.[18,20] Why is this? There are likely a multitude of reasons that are summarized in Fig. 3. However, in order to do better there is a need for the growing work force to have access to the specialized health services that will optimize health management as disease becomes increasingly multisystemic in the aging patients with ACHD. This is underscored by the observation that in geriatric CHD populations, it has been shown that the predictors of death relate to cancer, dementia, and kidney disease, with a decreasing emphasis on heart disease itself and with an amplification of acquired cardiovascular complications.[4]

In summary, as demonstrated with population data in Canada, the United States, and Europe, there is no longer any question that CHD is a lifespan condition, with adults comprising the fastest growing segment of this population. The observation that there is a predominance of women of reproductive age compared with men compels us to consider the need for specialized obstetric care as reproductive fitness in women with CHD evolves. This life stage is one of several that patients with ACHD will experience as longevity continues to increase underscoring the need for a novel life course epidemiology construct that accounts for determinants and processes of health more comprehensively. As we seek not just to improve mortality but change outcomes with the goal of improving quality of life, there is an opportunity to address the multisystemic nature of ACHD, seeking to implement interdisciplinary care to tackle potentially reversible multisystemic cardiovascular and noncardiovascular disease complications.

ADULT CONGENITAL HEART DISEASE—A MULTISYSTEM CONDITION

Why Is the Paradigm Shifting?

ACHD is a multisystem disease. Both cardiac and noncardiovascular complications have contributed to this multisystem disease and will require lifelong surveillance (Box 1).[21] In a single center trial of 6969 adult patients with CHD who were followed-up between 1991 and 2013, 524

Box 1
Cardiovascular and noncardiovascular complications

- Cardiovascular Complications
  - Heart failure
  - Arrhythmia
  - Residual valvular or shunt abnormalities
  - Prosthetic materials
- Noncardiovascular Complications
  - Endocrine
    - Bone health
    - Calcium hemostasis
    - Diabetes
    - Dyslipidemia
    - Metabolic syndrome
    - Obesity
    - Thyroid
  - Hematology
    - Anemia
    - Hyperuricemia
    - Iron deficiency
    - Secondary erythrocytosis
    - Thromboembolism
  - Immunology/Infectious Disease
    - Brain abscess
    - Endocarditis
    - Pneumonia
    - Protein-losing enteropathy
  - Liver
    - Cardiac cirrhosis
    - Congestive hepatopathy
    - Fontan-associated liver disease
  - Lung
    - Plastic bronchitis
    - Pulmonary hemorrhage
    - Pulmonary hypertension
    - Restrictive lung disease
  - Oncology
    - Age-appropriate cancer screening
    - Low-dose ionizing radiation and malignancy
    - Hepatocellular carcinoma
  - Brain
    - Neurocognitive deficits

- Depression
- Anxiety
- Renal
  - Cardiorenal syndrome
  - Chronic kidney disease
- Vascular
  - Aortopathy
  - Cerebrovascular disease
  - Endothelial dysfunction
  - Hypertension
  - Peripheral venous/arterial disease

patients died over a median follow-up of 9.1 years.[22] The leading cardiac causes of death were heart failure (42%) and sudden death (7%), whereas noncardiovascular causes were just as important, including pneumonia (10%) and cancer (6%).[22] Acquired cardiovascular risk factors such as diabetes, hypertension, and obesity will also factor into the outcome of older patients with CHD.[23] This section focuses on the multisystem disease that adults with CHD develop over the long term.

Cardiovascular Complications

Residual hemodynamic and electrophysiologic abnormalities play an important role in the cardiovascular outcome of adults with CHD. Nearly 25% of adults with CHD will develop heart failure at the age of 30 years.[24] A multitude of factors can lead to heart failure, including residual ventricular dysfunction, valvular disease, shunts, and arrhythmias. Heart failure with reduced ejection fraction is commonly seen in patients with a morphologic systemic right ventricle or a single ventricle after Fontan palliation, which is predisposed to cardiac dysfunction. Less attention has been paid to heart failure with preserved ejection fraction (HFpEF) in adults with CHD. Although more than 50% of general adult cardiology patients may be affected, HFpEF may be as prevalent in the ACHD population.[25] Fontan failure with preserved ejection fraction remains one of the most challenging types of heart failure to treat and is a marker for worse outcomes with transplantation.[26] There are opportunities to mitigate these cardiac complications through follow-up at ACHD centers, which has been shown to improve survival in this population.[19] It will be important to monitor for repeat intervention on residual hemodynamic lesions; approximately 20% of patients with CHD will require surgery after they turn adults.[27] The most common indication for reoperation is pulmonary valve replacement in the setting of tetralogy of Fallot (TOF). These operations are often being performed earlier before the onset of symptoms in order to prevent right heart failure and arrhythmia later in adulthood.

Arrhythmia is one of the most frequent indications for hospitalization and cause of death in adults with CHD.[28] More than 50% of adults with CHD are estimated to develop an atrial tachyarrhythmia during their lifetime.[28] There is a 2% to 5% incidence per decade of sudden cardiac death (SCD) in individuals with TOF and transposition of the great arteries after atrial switch.[28,29] Monitoring for arrhythmias and the use of invasive electrophysiologic procedures can be important in risk stratifying individuals with CHD who may be at risk for SCD. Although moderate and complex defects are more likely to suffer from cardiovascular complications, simple defects are not immune. The risk of endocarditis persists lifelong for all defects,[30] with even small unoperated ventricular septal defects having a risk of infective endocarditis (IE) that is 20 to 30 times that of the general population.[31] Guideline-based antibiotic prophylaxis remains important in the reduction of IE risk over a lifetime as well as a low threshold to obtain blood cultures in individuals with CHD before antibiotic treatment in the setting of fever of unknown origin. The importance of routine dental care and updated vaccinations are important preventative strategies.

Noncardiovascular Complications

Noncardiovascular complications are increasingly prevalent in adults with CHD with nearly all organ systems affected (**Fig. 4**). More than 40% to 50% of adults with CHD are noted to have abnormal renal and pulmonary function tests.[32,33] These noncardiac conditions have more than doubled in patients with CHD who have been hospitalized between 1998 and 2010.[34] Renal dysfunction, restrictive lung disease, anemia, and liver cirrhosis have all been associated with reduced survival in patients with CHD.[21] Both cardiac and noncardiac surgery can be affected by these noncardiac comorbidities with an increased likelihood of developing acute renal failure, pneumonia, and respiratory failure postoperatively.[35] Unique endocrine and immunologic complications are seen in individuals with CHD with genetic syndromes.[36] Patients with CHD with residual cyanosis are the most vulnerable to noncardiovascular complications affecting nearly all organ systems including unique complications from

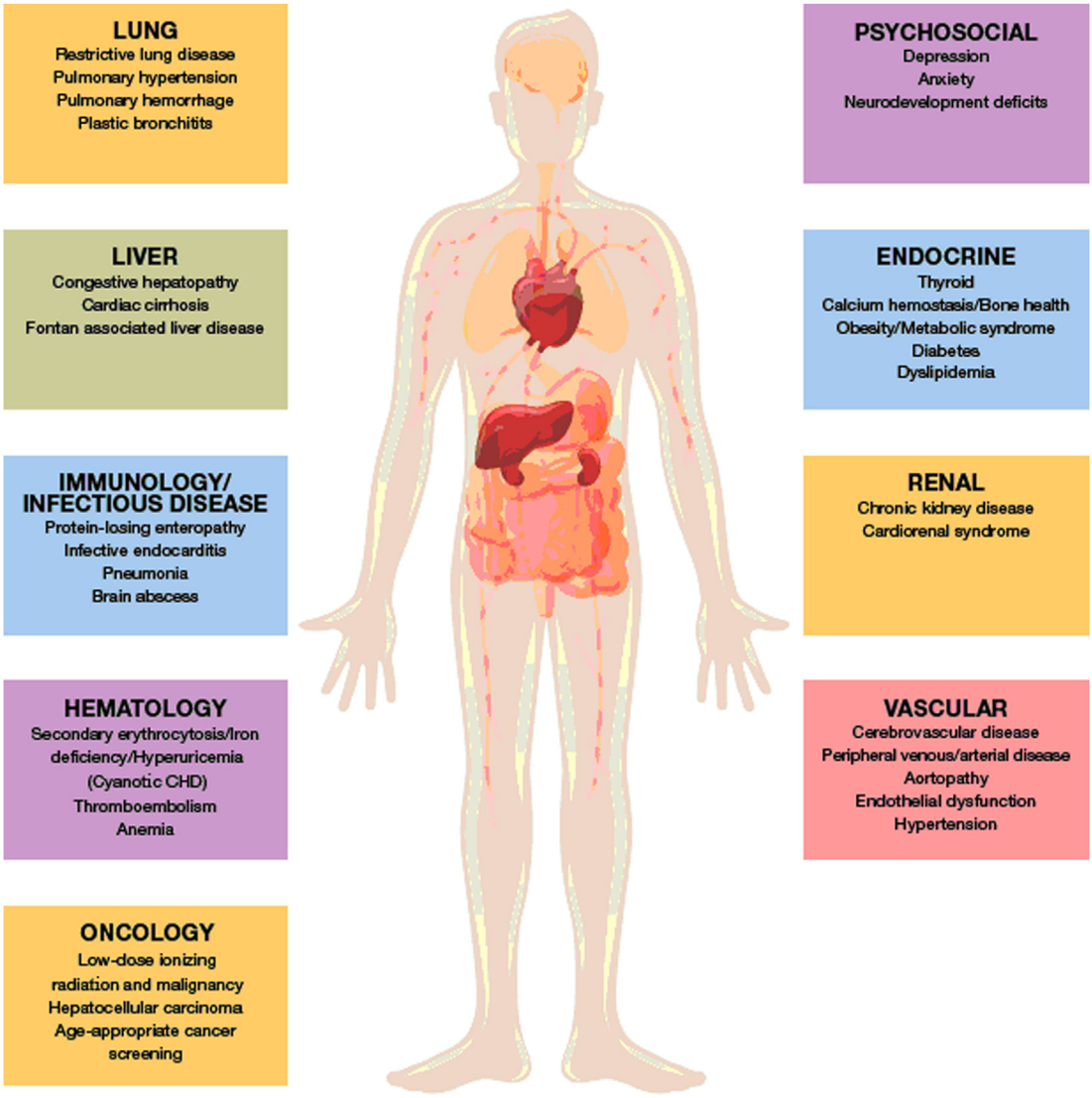

Fig. 4. Noncardiac complications in adults with congenital heart disease. (*Reprinted with permission from* Lui GK, Saidi A, Bhatt AB, Burchill LJ, Deen JF, et al. American Heart Association Adult Congenital Heart Disease Committee of the Council on Clinical C, Council on Cardiovascular Disease in the Y, Council on Cardiovascular R, Intervention, Council on Quality of C and Outcomes R. Diagnosis and Management of Noncardiac Complications in Adults With Congenital Heart Disease: A Scientific Statement From the American Heart Association. Circulation 2017;136:e348-e392 ©2017 American Heart Association, Inc.)

secondary erythrocytosis.[21] Cancer is the second leading cause of noncardiovascular death in adults with CHD.[22] The prevalence of cancer in adults with CHD in Québec is 1.6 to 2 times higher than that of the general population.[37] Risk factors likely include prior radiation exposure, genetic factors, and even unique CHD repairs such as the Fontan palliation, which has been associated with hepatocellular carcinoma.[21] Neurodevelopmental deficits increase in frequency and severity with CHD complexity. There is significant research on the prevalence and types of neurodevelopmental disabilities in children and young adolescents and a growing body of evidence on the impact of CHD on the brain of patients with ACHD.[38,39] Not only is there an increased incidence of stroke in patients with ACHD,[40] but recent evidence suggests the possibility of an accrued risk of dementia[41] potentially mitigated by the cumulative burden of vascular complications in the brain of patients with CHD.[38] In addition, more than one-third of adults with CHD have reported a mood or anxiety

disorder.[42] Both cognitive challenges and psychosocial distress can have profound effects on health care, education, employment, and overall quality of life of patients with CHD. Finally, as the number of adults with CHD older than 65 years increases, acquired cardiovascular diseases will have a significant impact; more than 80% of adults with CHD have more than one atherosclerotic cardiovascular risk factor.[43] Adult comorbidities such as diabetes, coronary artery disease, and hypertension will begin to shift how practitioners who once only focused on the late cardiac sequelae of CHD to managing a multisystem disease including both cardiovascular and noncardiovascular complications of adults with CHD.

In summary, the key to management of extracardiac complications is early detection, which requires an interdisciplinary team with expertise in disciplines such as hepatology, immunology, pulmonology, and nephrology as well as familiarity with complex CHD. The integration of mental health providers into ACHD care teams provide an opportunity for early identification and management of psychosocial distress. In order to improve the long-term outcome of this vulnerable patient population, understanding and managing noncardiovascular complications becomes as important as knowing their cardiac history. As these individuals grow in numbers and age, we need to identify preventative strategies with intervention at an earlier age to mitigate the development of later cardiovascular and noncardiovascular complications. Thus, processes of care become key drivers of the quality of the care that we can plan to deliver. This is especially important as we implement quality improvement (QI) initiatives in partnership with the growing number of care givers that the ACHD workforce mandates.

DELIVERING QUALITY IN ADULT CONGENITAL HEART DISEASE CARE—CAN WE DO IT?

How Do We Define Quality of Care for Patients with Adult Congenital Heart Disease?

Quality of care was defined by the Institute of Medicine (IOM) as"the degree to which health services for individuals and populations increase the likelihood of desired health outcomes and are consistent with current professional knowledge."[44] The basic tenets of delivering quality care from the IOM include that care should be safe, effective, patient centered, timely, efficient, and equitable.[45] Because these tenets were developed in the early 1990s, health care systems and providers have adapted practices to ensure these goals come to fruition. In earlier decades, quality care for CHD used to mean survival. However, with survival to adulthood approaching 90%, the focus of care has turned to addressing morbidity and long-term outcomes, patient experience, and quality of life. Thus, the focus of quality of care has also shifted to include these aspects of care beyond survival.

The foundational aspects of quality of care assessment are based in the models of Donabedian[46] and, more recently, Porter.[47] The Donabedian model frames quality of care into 3 specific areas: structure, process, and outcome. Structure involves the physical context of the health care system including who is seeing patient, in what types of locations, and with what equipment. Process includes the technical aspects of health care delivery including timing and types of testing, procedures and medications, and how patients flow through the system. Outcome includes both that of the patient and the health care system, including aspects such as mortality, clinical outcomes, costs, and efficiency. The Porter model adds on value-based health care and the idea that health care should be centered on outcomes that matter to patients and the costs to achieve those outcomes.

One way to bring these quality assessments into practice is to break them down to 3 components: quality measurement, quality reporting, and QI. Quality measurement is a retrospective approach to assess the quality of care delivered, which may include the proportion of women receiving recommended screening mammography or proportion of people offered flu shots each year. Quality reporting is the transparent approach to sharing quality measurement data to inform future efforts. QI is a prospective approach to improving care to individual patients and health care systems, which might include implementation of decision support tools for mammogram or flu shot reminders.

Importance of Quality of Care for Patients with Adult Congenital Heart Disease

In ACHD, developing consistent mechanisms to measure and improve quality of care is particularly important but also complicated. ACHD comprises patients not only with a great variety of underlying congenital heart conditions but also a variety of treatment strategies. Even for the same underlying conditions, treatment strategies may vary based on the era of birth and the available surgical intervention at that time or in that geographic area. For example, a patient born in 1990 with d-loop transposition of the great arteries may have undergone 1 of 2 surgeries depending on the surgical services available to them at the time. Similarly, a patient with tetralogy of Fallot born in the 1950s would likely have undergone a Blalock-Taussig shunt in infancy or

childhood followed by eventual intracardiac repair where a patient born in the 2000s would have had a single intracardiac repair in infancy.

Also contributing to the complexity of devising and delivering quality care to patients with ACHD is the large proportion of patients who have gaps in care or are lost to follow-up and the different types of providers who care for patients with ACHD in adulthood. More than 40% of patients with CHD have gaps in congenital cardiac care of greater than 3 years[48] and these lapses in care start as early as childhood.[49] These lapses in care can change the trajectory of the condition, as those with gaps are more likely to require urgent interventions on return to congenital heart care.[50] Furthermore, as patients with CHD transition to adulthood, many are unable or unaware of the need to find a cardiologist specializing in congenital heart care. Many patients end up in general cardiology care, primary care, or no medical care at all, and the specific needs for the patient with ACHD may not be recognized or met. This is most evident in the care of patients with ACHD with underlying complex CHD where maintaining specialized ACHD care has a documented survival benefit.[19]

The tremendous underlying variation in disease, treatment, and care provision, providing a foundational way to measure and improve quality in this population, is of great importance. Specific quality measures and reporting can also provide consistency in data collection for process and outcomes that are critical for further research and data acquisition. This information can be used to design studies and refine care processes for improvement in ACHD care as well as provide feedback to improve care in pediatrics.

Quality Initiatives in Adult Congenital Heart Disease Care

With the recognition of the need and importance for quality assessment in ACHD care, there have been multiple initiatives to measure and improve quality of care over the past few decades. A key feature of quality of care planning is that information must be easy to spread to many different types of providers across the community. These efforts started with the development of guidelines for care for the patients with ACHD published in Canada[51] and Europe[52] that are being updated. They have been updated in 2018 in the United States to include information about clinical management for the different CHD conditions as well as different types of repairs.[1] Some of the guidelines also include components related to specialized ACHD program design and support. Additional papers have been published evaluating guideline adherence in different countries. Although guidelines do not substitute for quality measures, they do provide a foundation for consistency of care and for the design of quality measures and QI activities. The observation that adherence to clinical practice guidelines is highly variable for patients with ACHD underscores the heterogeneity of the patient population and the need for more individualized approaches to care as data continue to evolve.[53,54]

Based on the available guidelines at the time, ambulatory quality measures for 6 ACHD conditions were developed in 2013.[55] The conditions included atrial septal defect, coarctation of the aorta, d-loop transposition of the great arteries, tetralogy of Fallot, Fontan procedure, and Eisenmenger syndrome. These measures were developed using the RAND-modified Delphi method[56] and resulted in 55 total measures for ambulatory care across the 6 conditions. Subsequent to this, additional studies have been performed evaluating electronic data collection of measures and implementation of care processes across populations. The measures can also serve as a foundation to design and implement QI projects.

In addition to the quality assessment for processes and outcomes of clinical care described, there have been efforts in ACHD to ensure consistency of training and resources across ACHD providers and programs. Until 2012, there were no criteria for specialized adult congenital heart disease training for health care providers; fellowships existed but were not standardized across programs for content, duration, or competencies. After significant effort by ACHD providers, the American Board of Medical Specialties approved adult congenital heart disease as separate board-certified medical specialty in 2012 and the first board examination was administered in 2015.[57] This has resulted in hundreds of board-certified ACHD specialists across the country and multiple consistent fellowship training programs. In addition to specialty certification for individuals, the Adult Congenital Heart Association, the largest ACHD patient advocacy organization in the United States, worked with its medical advisory board to develop accreditation criteria for ACHD programs across the United States to improve quality and consistency of care across programs and across the country.[58] There are currently 35 accredited programs across 23 states.

In summary, quality assessment has many components including measurement, reporting, and improvement. Efforts in multiple types of quality

of care have been developed in ACHD, resulting in quality measures to use for assessment and improvement initiatives as well as building consistency in training and resources across providers and programs. These efforts provide a foundation but will need to go further to improve processes and outcomes in all aspects of care including inpatient, outpatient, and procedures.

SUMMARY AND FUTURE DIRECTIONS

The demographic shifts in the ACHD population are rapidly changing in the United States, with a predominance of adults compared with children (see **Fig. 1**; see **Table 1**). This mandates the need for more adult cardiology care providers with expertise in CHD, a condition considered largely in the prevue of pediatric cardiologist just more than 30 years ago. There is predominance of women (see **Fig. 2**) where sex and gender determinants of outcomes will need to be accounted for in addition to the growing expertise that will be needed in specialized obstetric care. It is becoming increasingly evident that ACHD is a multisystem condition with a myriad of cardiovascular complications but also a growing body of evidence that support the clinical observations of complications distal to the heart including, metabolic complications, liver and kidney disease, vascular health, complications along the heart-brain axis, and cancer (see **Fig. 4**; see **Box 1**). Without a doubt managing multisystem disease will require the close collaboration of interdisciplinary teams where there is ongoing communication not only between pediatric and adult providers but also between adult subspecialties in cardiovascular and other medical subspecialties. Cost containment for populations with chronic, lifelong morbidity challenges our ability to sustain delivery of high-quality care underscoring the need for process-related measures of care quality such as that have been developed for ACHD outpatient management. Surveillance that is patient centered and sufficiently standardized to ensure that care is delivered commensurate with guideline recommendations is critical to prevention of complications but also challenging for a group of patients who are largely active and mobile. This requires organization of care at a systems level in a way that is well aligned with country-specific health insurance models.

As the body of evidence grows that CHD is a dynamic life-long complex series of pathophysiological disturbances, so too the need for shift in conceptual models that will underpin future research directions. As illustrated in **Fig. 3**, the life stages that a patient with CHD lives through require numerous transitions and turning points in their health management journey. Determinants of health are complex as are the processes of health development for this patient population. Ultimately, the goals of the workforce are to improve life-expectancy and functional capacity, reduce disability, and promote social integration of people with CHD. Increasingly, our interest is not only in improving disease trajectories but also in maximizing wellness trajectories and minimizing biological aging trajectories. This serves as a health services policy framework that moves toward the precision delivery of health services to provide cost-effective life course health management, organized around developmental time frames that will promote the longitudinal integration of health services across the lifespan.[59]

DISCLOSURE

The authors declare no conflict of interest.

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