Adult Congenital Heart Disease: A Growing Epidemic

Pablo Ávila, MD, Lise-Andrée Mercier, MD, Annie Dore, MD, François Marcotte, MD, François-Pierre Mongeon, MD, MS, Reda Ibrahim, MD, Anita Asgar, MD, Joaquim Miro, MD, Gregor Andelfinger, MD, Blandine Mondésert, MD, Pierre de Guise, MD, Nancy Poirier, MD, and Paul Khairy, MD, PhD

Montreal Heart Institute Adult Congenital Center, Université de Montréal, Montreal, Quebec, Canada

ABSTRACT

Medical and surgical breakthroughs in the care of children born with heart defects have generated a growing population of adult survivors and spawned a new subspecialty of cardiology: adult congenital heart disease. The prevalence of adult congenital heart disease is escalating at a rampant rate, outpacing the relatively static prevalence of pediatric congenital heart disease, because adults now surpass children in numbers by a ratio of 2:1. As such, congenital heart disease can no longer be considered primarily a pediatric specialty. Most congenital heart defects are not curable and require lifelong specialized care. Health care systems worldwide are challenged to meet the unique needs of this increasingly complex patient population, including the development of supraregional centres of excellence to provide comprehensive and multidisciplinary specialized care. In this review, we explore the incidence and prevalence of congenital heart disease and their changing patterns, address organization and delivery of care, highlight the importance of appropriate training and dedicated research, summarize the high burden of health care resource utilization, and provide an overview of common issues encountered in adults with congenital heart disease.

Congenital heart malformations are the most common birth defects and the leading cause of infant mortality. Although several studies have attempted to quantify the incidence of congenital heart disease, generating accurate estimates is complicated by nonstandardized definitions of qualifying pathologies, nonuniform screening methods, and geographic variability. Notwithstanding these limitations, recent estimates suggest that the incidence of congenital heart disease (excluding patent foramen ovale and bicuspid aortic valve) is in the order of 7 to 10 per 1000 live births, with moderate and severe forms afflicting 3 per 1000 newborns. Considering that there are approximately 138 million live births per year worldwide yields an estimated 1 million newborns with congenital heart disease every year. These rates have remained relatively constant since 2011.

In developed countries, a shift toward a greater proportion of simpler defects has been reported, which might reflect enhanced sensitivity in detecting milder forms of congenital heart disease combined with a reduction in the incidence of newborns with the most severe defects because of pregnancy termination on prenatal detection.
Prevalence of Congenital Heart Disease

Most patients born with moderate or severe forms of congenital heart disease in the 1950s and earlier died before reaching adulthood, with the highest mortality during the first week of life. Remarkable advances in diagnostic methods, medical management, interventional techniques, congenital heart disease surgery, and perioperative care have led to historical shifts in population demographic characteristics, and adults now outnumber children with congenital heart defects by a ratio of 2:1. Improved survival has been driven by a reduction in mortality in the youngest age strata, particularly in those with severe forms of congenital heart disease (Fig. 1). From 1987 to 2005, a > 30% reduction in mortality was reported among patients with congenital heart disease at large, with a 67% mortality reduction in children with complex disease. More than 85% of children born with heart defects are now expected to survive to adulthood and continue to thrive. Consequently, the population with congenital heart disease is aging. For example, the median age of patients with severe defects was 11 years in 1985 compared with 17 years in 2000. In 2000, the median age of adults with congenital heart disease was 40 years, with a median age of 29 in those with severe defects. From 1987 to 2005, the median age at death increased by 15 years, from 60 to 75 years, and the median age at death in those with severe defects increased from 2 to 23 years.

In a manner similar to incidence, estimating the prevalence of congenital heart disease is fraught with assumptions and subject to limitations. Prevalence is defined as the number of living subjects with a particular disease during a specific time frame. Birth prevalence underestimates the true burden of congenital heart disease by not capturing those in whom the diagnosis is established afterward. Prevalence estimated at a later time excludes those who died before the assessment, thereby differentially underrepresenting higher-fatality lesions, and potentially omitting defects that resolve spontaneously. For example, up to two-thirds of ventricular septal defects diagnosed at birth spontaneously close within the first years of life and will not be present in adulthood, whereas other forms of congenital heart disease, such as atrial septal defects, Ebstein anomaly, and congenitally corrected transposition of the great arteries might go unrecognized until adulthood. Furthermore, prevalence estimates might not consider other factors that affect global disease epidemiology, such as migration and population mobility.

A Quebec population-based study estimated that, in the year 2010, the prevalence of congenital heart disease in adults (18 years of age and older) was 6.1 per 1000. Similar rates were projected in a recent systematic review. Extrapolating this statistic to the general population, it might be estimated that there are > 100,000 adults with congenital heart disease in Canada, 1 million in the United States, and 1.8 million in Europe. It is also noteworthy that the prevalence of complex congenital heart disease in adults has been steadily increasing (eg, by 85% from 1985 to 2000), unlike the relatively stable prevalence of complex congenital heart disease in children (Fig. 2). By 2010, adults accounted for 60% of patients with complex congenital heart disease.

Organization and Delivery of Care

Although the provision of care for children with congenital heart disease is well established in developed countries, clinical services for adults are comparatively scarce. To attend to the progressive increase in the number of adults with congenital heart disease and the increasing complexity of their pathologies, specialized adult congenital heart centres have emerged in several countries, with increasing volume of activities (Fig. 3).

Canada assumed a leadership role in establishing recommendations for optimal organization and delivery of care. In 1991, the Canadian Adult Congenital Heart (CACH) network was established as the first national society dedicated to the care of adults with congenital heart disease (www.cachnet.ca). The Canadian Cardiovascular Society, in collaboration with the CACH network, formed a panel of experts in 1996 to discuss the needs of adults with congenital heart disease and provide recommendations for referral to specialized centres. The first management guidelines were presented at the Canadian Cardiovascular Congress in Montreal in 1996, published in the Canadian Journal of Cardiology in 1998, and subsequently updated in 2001 and 2009. Similar proposals emerged from the 32nd Bethesda Conference in 2000, which focused on the need for health care professionals, patients, and governing agencies to develop strategic plans to improve access to and delivery of care for the adult with congenital heart disease. This call to action was followed by more specific recommendations from the American College of Cardiology/American Heart Association and the European Society of Cardiology (ESC). Current management guidelines suggest that approximately half of the adult population with congenital heart disease stand to benefit from specialized care within adult congenital heart centres. Such care is generally recommended for the initial assessment of adults with known or suspected congenital heart disease, follow-up of patients with moderate and severe lesions, cardiac surgical and nonsurgical interventions, and risk assessment and support for pregnancy and noncardiac surgery.
Recommended personnel and services for regional adult congenital centres are summarized in Table 1.\textsuperscript{25} In short, centralized care within regional centres should be provided by a multidisciplinary team comprised of cardiologists with expertise in adult congenital heart disease, including imaging, arrhythmia management,\textsuperscript{28} interventional cardiology, and high-risk obstetrics, congenital heart surgeons, perioperative care, and psychosocial support.\textsuperscript{17,25,26} Estimates regarding the ideal number of regional adult congenital centres have varied between 1 per 2-10 million inhabitants,\textsuperscript{17,21} which translates to up to 17 specialized centres in Canada, 150 in the United States, and 365 in Europe.\textsuperscript{29} This figure balances factors such as ease of access and centralization of resources, considering that improved outcomes are proportional to volume of activity and experience.\textsuperscript{30} Although the recommended number of specialized centres has been achieved in Canada (ie, 17 centres are registered within the CACH network), most countries in the developed world fall short of this target. For example, in the United States, 108

Figure 2. Number and proportion of adults and children with (A) all forms of congenital heart disease (CHD), and (B) severe CHD in 1985, 1990, 1995, and 2000. Reproduced from Marelli et al.\textsuperscript{13}

Figure 3. Number of patients followed at the Montreal Heart Institute Adult Congenital Centre over time. A >7-fold increase in the number of adults with congenital heart disease was observed over a 15-year period.
exceed published recommendations. Specialized centres and that waiting times to access services <
portion of qualifying patients who actually receive specialized no guarantee for optimal care. A larger issue that plagues the Congenital Heart Association Web site (www.achaheart.org)
self-declared adult congenital heart centres with variable volumes of activity and availability of resources are listed on the American Congenital Heart Association Web site (www.achaheart.org).
An adequate ratio of specialized adult congenital centres is no guarantee for optimal care. A larger issue that plagues the field of congenital heart disease is the relatively small proportion of qualifying patients who actually receive specialized adult-oriented care. For example, a Canadian study found that < 25% of adults with congenital heart disease are followed by specialized centres and that waiting times to access services exceed published recommendations. These sobering statistics have prompted numerous educational initiatives to raise awareness about the need for lifelong care and have incited research studies into factors associated with gaps in care and impediments to long-term follow-up. Common barriers to transfer from pediatric-to adult-oriented health care include inadequate knowledge regarding the need for such care, a dearth of accessible competent adult care providers, and emotional attachment of patients and families to their pediatric caregivers and vice-versa. Lending further credence to the notion that specialized adult congenital centres with multidisciplinary expertise offer the best prospects for improving outcomes, a recent population-based study concluded that referral to such a centre is associated with a significant reduction in all-cause mortality (Fig. 4).
Training and Research
An important element in optimizing outcomes is the development of training programs to meet workforce requirements and provide qualified, consistent, and comprehensive care. Exposure of adult cardiology trainees to adult congenital heart disease didactic and clinical experience varies widely, reflecting the fact that few programs offer advanced training in this sector of cardiology. Surveys suggest that only approximately 25% of cardiologists who care for adults with congenital heart disease have received formal training in this discipline. The United States is the first country to officially recognize the legitimacy of adult congenital heart disease as a separate subspecialty of cardiology. In September 2012, the American Board of Medical Subspecialties approved a specialized certification examination via the American Board of Internal Medicine. The first examination is planned for 2015. Pediatric or adult cardiologists require a minimum 2-year fellowship within recognized adult congenital training centres to qualify. The ESC, with its working group on grown-up congenital heart disease, is poised to follow suit. Canada is currently contemplating recognizing adult congenital heart disease as an area of focused competence via a Diploma of the Royal College of Physicians and Surgeons of Canada (DRCPSC). While details remain to be determined, a Diploma of the Royal College of Physicians and Surgeons of Canada typically requires 1-2 years of additional training by Royal College-accredited programs and is competency-based, with assessment through a summative portfolio. Another critically important element required to advance the care of adults with congenital heart disease is well-designed research. Over the past decade, significant strides have been achieved in coordinating research efforts and creating dedicated infrastructures. A critical mass of patients and investigators has enabled a new era of multicentre research with more exacting standards. Several networks and organizations have emerged, some focused on multicentre research as their main objective, with others providing a supportive environment to facilitate research. These include, but are not limited to, the International Society for Adult Congenital Heart Disease (www.isachd.org), CACH network, ESC (www.escardio.org), Japanese Circulation Society (www.j-circ.or.jp/english), Society of Thoracic Surgeons (www.sts.org), Congenital Heart Surgeons’ Society (www.chss.org), and the Adult Congenital Heart Association. In the Netherlands, the Interuniversity Cardiology Institute and Heart Foundation developed a highly productive national registry and DNA bank of patients with congenital heart disease, named CONGenital CORvita (CONCOR; www.concor.net/en). In North America, an Alliance for Adult Research in Congenital Cardiology was founded in 2006 to foster collaborative research, advance knowledge, and improve outcomes (www.aarcc.net).

### Table 1. Personnel and services recommended for regional ACHD centres

<table>
<thead>
<tr>
<th>Type of Service</th>
<th>Personnel/Resources</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiologist specializing in ACHD</td>
<td>One or several 24/7</td>
</tr>
<tr>
<td>Congenital cardiac surgeon</td>
<td>Two or several 24/7</td>
</tr>
<tr>
<td>Nurse/physician assistant/nurse practitioner</td>
<td>One or several</td>
</tr>
<tr>
<td>Cardiac anaesthetologist</td>
<td>Several 24/7</td>
</tr>
<tr>
<td>Echocardiography*</td>
<td>Two or several 24/7</td>
</tr>
<tr>
<td>Diagnostic catheterization*</td>
<td>Yes, 24/7</td>
</tr>
<tr>
<td>Noncoronary interventional catheterization*</td>
<td>Yes, 24/7</td>
</tr>
<tr>
<td>Electrophysiology/pacing/ICD implantation*</td>
<td>One or several</td>
</tr>
<tr>
<td>Exercise testing</td>
<td>*Echocardiography</td>
</tr>
<tr>
<td>Cardiac imaging/radiology*</td>
<td>*CT scanning</td>
</tr>
<tr>
<td>Multidisciplinary teams</td>
<td>*Nuclear medicine</td>
</tr>
<tr>
<td>*High-risk obstetrics</td>
<td>*Pulmonary hypertension</td>
</tr>
<tr>
<td>*Heart failure/transplant</td>
<td>*Genetics</td>
</tr>
<tr>
<td>*Neurology</td>
<td>*Nephrology</td>
</tr>
<tr>
<td>*Cardiac pathology</td>
<td>*Rehabilitation services</td>
</tr>
<tr>
<td>*Social services</td>
<td>*Vocational services</td>
</tr>
<tr>
<td>*Financial counsellors</td>
<td>*Cardiac MRI</td>
</tr>
<tr>
<td>Information technology</td>
<td>*MRI</td>
</tr>
<tr>
<td>Database collection</td>
<td>*Nuclear medicine</td>
</tr>
<tr>
<td>Database support</td>
<td>*PACU</td>
</tr>
<tr>
<td>Quality assessment</td>
<td>*Electrophysiology</td>
</tr>
<tr>
<td>review/protocols</td>
<td>*Catheter imaging</td>
</tr>
</tbody>
</table>

24/7, availability 24 hours per day, 7 days per week; ACHD, adult congenital heart disease; CT, computed tomography; ICD, implantable cardioverter defibrillator; MRI, magnetic resonance imaging; TEE, transesophageal echocardiography.

*These modalities must be supervised/operated and interpreted by physicians with expertise and training in congenital heart disease.

Reproduced from Warnes et al. The United States is the first country to officially recognize the legitimacy of adult congenital heart disease as a separate subspecialty of cardiology. An American Board of Medical Subspecialties approved a specialized certification examination via the American Board of Internal Medicine. The first examination is planned for 2015. Pediatric or adult cardiologists require a minimum 2-year fellowship within recognized adult congenital training centres to qualify. The ESC, with its working group on grown-up congenital heart disease, is poised to follow suit. Canada is currently contemplating recognizing adult congenital heart disease as an area of focused competence via a Diploma of the Royal College of Physicians and Surgeons of Canada (DRCPSC). While details remain to be determined, a Diploma of the Royal College of Physicians and Surgeons of Canada typically requires 1-2 years of additional training by Royal College-accredited programs and is competency-based, with assessment through a summative portfolio.

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admissions, and cardiac interventions are several-fold greater than in the general population. Models of health-related costs and outcomes in adults with congenital heart disease provide economic justification for expensive interventions on the basis of striking benefits on quality-adjusted life years.

Overview of Common Issues

Common complications that contribute to the growing epidemic of adults with congenital heart disease include arrhythmias, heart failure, pulmonary hypertension, endocarditis, pregnancy-related issues, and cardiac interventions.

Arrhythmias

Arrhythmias are the most common long-term complication in adults with congenital heart disease, the leading source of morbidity and hospital admissions, and a major cause of mortality. All forms of tachy- and bradyarrhythmias might be encountered. Figure 6 schematically summarizes factors predisposing to arrhythmias. The prevalence of tachyarrhythmias generally correlates with increasing age and complexity of the congenital heart lesion (Fig. 7).

Heart failure

Mechanisms for heart failure in adults with congenital heart disease might involve circulatory overload, ventricular volume and/or pressure overload, inability of a systemic morphologic right ventricle or single ventricle circulation to adequately meet metabolic needs, unequal flow distributions to lung fields, variable pulmonary vascular integrity, and abnormalities of myocardial tissue architecture, electrical conduction, and blood supply. As such, management guidelines for acquired heart disease cannot be extrapolated to the complex forms of heart failure encountered in adults with congenital heart disease. Despite the heterogeneity and imprecisions, on the whole, heart failure appears to be increasing in prevalence as the population ages. It is among the most common causes of hospitalization (see Fig. 5), with some studies suggesting that it has surpassed sudden cardiac death as the leading cause of mortality. The incidence of systolic systemic ventricular dysfunction is highest in those with cyanotic lesions, systemic right ventricles, and univentricular hearts. In some forms of congenital heart disease, diastolic systemic ventricular dysfunction has been correlated with arrhythmias and mortality. Even in the absence of overt heart failure as classically defined, some degree of exercise intolerance is documented in up to one-third. When contrasted against quantified exercise capacity, functional symptoms are notoriously underreported as a result of lifelong adaptation to a chronic disorder. Moreover, in an increasing number of adults with congenital heart disease and end-stage circulatory failure in whom cardiac transplantation is not a viable option, end-of-life issues and palliative care assume a prominent role.

Pulmonary hypertension

Any nonrestrictive communication causing left-to-right shunting with increased pulmonary blood flow might produce pathological changes to the pulmonary vascular bed,
including vasoconstriction, inflammation, cell proliferation, and vascular remodelling, resulting in increased pulmonary vascular resistance.\textsuperscript{69,70} Pulmonary arterial hypertension is prevalent in 4%-10% of adults with congenital heart disease and is associated with exercise intolerance and heart failure.\textsuperscript{69,71} At the extreme end of the spectrum, Eisenmenger syndrome is a multisystemic disorder associated with a 10- to 12-fold increase in mortality in those with complex congenital heart disease.\textsuperscript{69,72-77} If managed appropriately, many such patients survive past the third or fourth decade of life.\textsuperscript{72} In developed countries, earlier recognition and closure of intracardiac shunts has resulted in a reduction in the prevalence of Eisenmenger syndrome.\textsuperscript{78} Nevertheless, pulmonary hypertension persists in 3%-13% of patients after septal defect closure\textsuperscript{71,79} and implies a worse prognosis.\textsuperscript{80}

**Endocarditis**

The incidence of endocarditis (approximately 1.1 per 1000 patient-years) in adults with congenital heart disease is 20-fold greater than in the general population,\textsuperscript{81,82} with a 12% recurrence rate.\textsuperscript{81,83} Factors associated with endocarditis include the type of congenital heart defect, presence of intracardiac shunts has resulted in a reduction in the prevalence of Eisenmenger syndrome.\textsuperscript{78} Nevertheless, pulmonary hypertension persists in 3%-13% of patients after septal defect closure\textsuperscript{71,79} and implies a worse prognosis.\textsuperscript{80}

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multiple defects, and male sex.81 Systemic embolization, heart failure, and valvular regurgitation are common complications.84 Surgery is required in up to one-third of patients.84,85 Improvements in antibiotic therapy and earlier surgical management have resulted in lower rates of complications and mortality over the past decades, with current in-hospital mortality ranging from 4% to 9%.81,83,84 Educational counselling, including hygienic measures and antibiotic prophylaxis when indicated, should be incorporated in the care of adults with congenital heart disease.

**Pregnancy**

When considered collectively, congenital malformations are the most prevalent form of heart disease in pregnant women86,87 and the leading cardiac cause of maternal morbidity and mortality in developed countries.25 Pregnancy is the most common noncardiac cause of hospitalization in women with congenital heart disease.45,46 Congenital heart disease carries increased maternal and fetal risks, particularly in the setting of pulmonary arterial hypertension, subaortic or subpulmonary ventricular dysfunction, left-sided valvular obstruction, un repaired aortic coarctation, or aortic root dilatation in association with other anomalies such as a bicuspid aortic valve, Marfan, or Turner syndrome.86-89 Heart failure and arrhythmias are the most common maternal complications, whereas frequent neonatal adverse outcomes include small for gestational age birth weight, respiratory distress, and intraventricular cerebral hemorrhage.90 Women with moderate and complex forms of congenital heart disease should be managed by a multidisciplinary team with appropriate expertise and delivery should take place in specialized centres.25-27,91

**Congenital cardiac surgeries and interventions**

Surgical and catheter procedures are on the increase in adults with congenital heart disease.92 Percutaneous devices are increasingly used to close septal defects, abnormal vascular connections, leaks across conduits and baffles, and pseudoaneurysms; covered stents have enhanced the safety with which aortic coarctation, conduit stenosis, and branch pulmonary stenosis can be treated; and percutaneous valves have broadened the range of therapeutic options in selected patients.93 Cardiovascular surgery is also increasingly performed in adults with congenital heart disease to rectify residual or progressive lesions in the setting of previous reparative surgery, address previously unoperated disease, palliate nonreparable defects, and transplant failing hearts.94 Complications after surgery occur in 15%-25% of patients and include arrhythmias, low cardiac output, stroke, bleeds, and renal failure. Perioperative mortality risk depends on numerous factors but is generally within the 3%-8% range.95-98

**Conclusion**

Adult congenital heart disease is the most rapidly growing sector of cardiology today, with a patient population that is swiftly increasing in size and complexity. Unique issues, challenges, and considerations are involved in managing adults...
with congenital heart disease, including arrhythmias, heart failure, pulmonary hypertension, endocarditis, pregnancy, interventions, and surgery. An increasing body of evidence suggests that outcomes can be improved by referral to specialized centres with multidisciplinary teams dedicated to clinical care, training, and research in adult congenital heart disease.

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**Disclosures**

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