Congenital Heart Disease Epidemiology in the United States
Blindly Feeling for the Charging Elephant

Congenital heart disease (CHD) is the most common of all congenital defects, affecting nearly 1% of live births. Creative operative and catheter-based interventions have facilitated survival to adulthood with nearly all defects, setting the stage for a rapidly growing population of adolescents and adults with CHD. Although the field of adult CHD (ACHD) remains in its infancy, it is important to acknowledge the tremendous achievements in the United States and abroad over the last decades. ACHD publications have increased almost exponentially (Figure). The first American College of Cardiology/American Heart Association joint guidelines on the diagnosis and management of ACHD were published in 2008 with a revision anticipated soon; subspecialty status in cardiology was granted by the American Board of Medical Specialties in 2012 with the first qualifying examination offered in the fall of 2015, and the first class of American Council for Graduate Medical Education–approved fellowship training begins July 2016. Multi-center and multinational research collaborations have formed, including the Alliance for Adult Research in Congenital Cardiology, as have patient/provider collaborative organizations such as the Adult Congenital Heart Association and the International Society of Adult Congenital Heart Disease. Funding for research has also improved, led by the US Department of Defense, the American Heart Association, and the National Heart, Lung, and Blood Institute.

Despite tremendous progress, obstacles to the appropriate and sustained growth of this field remain numerous in the United States. Gilboa and colleagues remind us of one of these major limitations in this issue of Circulation. Our current estimate of the number of patients with CHD in the United States continues to be based on extrapolations from the population of Quebec, Canada. Although Marelli and colleagues have published several seminal papers using the data from this well-characterized cohort of patients, it remains uncertain just how closely Canadian data parallel our own population. Canada has a lower infant mortality rate and a smaller population of obese patients, and the United States has 5.7-fold more blacks and 17.4-fold more Hispanics. Socioeconomic conditions, genetic backgrounds, and environmental exposures can all affect maternal and fetal health, thus influencing birth rates of CHD. Canadians enjoy universal access to health care, whereas access in the United States is too often based on socioeconomic conditions. Changes to our healthcare system have recently improved access for the indigent population, although evidence suggests that not everyone has benefited. Patients with CHD in the United States are particularly likely to experience a loss to follow-up in their late teens to early 20s, which appears to coincide with loss of insurance coverage and its potentially detrimental effects on overall health. Healthcare system changes may help curtail this problem, but the United States is poorly equipped to handle the number of ACHD patients who need care. Certain parts of the United States have also experienced dramatic
and unexpected demographic changes over the past few decades that provide additional unique challenges to the healthcare infrastructure.\textsuperscript{11}

It is becoming clear that the best way to understand the impact of CHD in the United States is to establish a national registry. Although this has been actively discussed in the community of physicians caring for these patients for several decades, a small step in this direction only recently followed the passage of the Congenital Heart Futures Act in 2010. With this appropriated funding, the US Centers for Disease Control and Prevention established a pilot study of 3 US centers to begin tracking CHD prevalence in their respective states. This group recently expanded to 5 centers that are using novel techniques to link multiple administrative and medical data sets to gain greater insight into the US CHD population. Until these data are fully collected, interpreted, and disseminated, large provincial and national registries outside the United States remain our most appropriate reference points. We need a much greater effort in the United States.

As the strongest current evidence, the findings of Gilboa and colleagues\textsuperscript{2} are eye-opening: 1.4 million adults with CHD were living in the United States in 2010, with 160,000 having severe disease. Those of us taking care of patients with CHD expected this growth in the ACHD population. The number of adults with CHD continues to outnumber children with this problem. Prior estimates derived from the Quebec data suggested a US ACHD population of \( \approx 850,000 \) in 2000.\textsuperscript{12} This population appears to be growing at a rate of \( \approx 40,000 \) to 50,000 new ACHD patients per year. Thus, the figure of 1.4 million adults with CHD in 2010 should have been expected. The 32nd Bethesda Conference on ACHD care in 2000 divided CHD into mild, moderate, and severe categories, with the last category contributing 14\% of the total.\textsuperscript{13} Thus, the estimated number of 160,000 adults with severe disease is also about the number expected. It appears, however, that despite these anticipated findings, we continue to underestimate the magnitude of the problem and the amount of resources necessary to appropriately manage it.

More than a third of ACHD patients are currently at least 45 years of age.\textsuperscript{2} The American Heart Association Council on Clinical Cardiology recently tabulated the multitude of medical issues attendant with becoming an older adult with CHD,\textsuperscript{14} with complex cardiac anatomy being only a single facet. Complex cardiac anatomy contributes to the well-known, costly comorbidities of heart failure, arrhythmias, and pulmonary hypertension. However, beyond these factors are frequently encountered neurological/psychological issues, problems with insurability and employability, pulmonary limitations, hepatic abnormalities, renal dysfunction, hematologic problems, endocrinopathies, and the challenging issues of pregnancy and appropriate contraceptive counseling and gynecological care.\textsuperscript{15} These burdens are in addition to the standard, guideline-based, age-appropriate screening and preventive care, which become increasingly important as the population ages. Practicing ACHD medicine thus requires a multifaceted skill set from a

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\caption{Trends in adult congenital heart disease (ACHD).}
\begin{flushleft}
PubMed cited articles directly relating to adults with CHD over 4-year intervals since 1980 (blue) and the estimated number of ACHD patients (from program survey by the Adult Congenital Heart Association and the International Society of Adult Congenital Heart Disease) seen at programs in the United States that self-report as providing specialized care to adults with congenital disease (red) during 4 different years since 2005.
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highly motivated and dedicated group of people with the ability to work in concert with a variety of different specialties familiar with the various unique CHD sequelae. Caring for these patients is a team sport.

The costs of inpatient care of ACHD appear to be increasing considerably, with hospital readmission becoming a progressively larger component. Care for expensive complications such as heart failure remains non-standardized, with little in the way of published studies in the ACHD population to reference in making evidence-based therapeutic decisions. Frustratingly, patients are all too often referred to specialty ACHD care late in the game when their condition has deteriorated beyond the point at which simple, or even complicated medical options (eg, pulmonary hypertension therapy), catheter-based interventions (ie, defect closure, percutaneous valve replacement, or electrophysiological treatment), palliative or curative surgical repair considerations, or advanced heart failure therapies, including organ transplantation, are even possible. The best way to prevent extensive use of hospital resources for often futile end-of-life care is to identify, treat, and, we hope, prevent many of the treatable complications in the outpatient setting before the wheels fall off.

The Adult Congenital Heart Association/International Society of Adult Congenital Heart Disease informally surveys ACHD programs in the United States on a yearly basis to assess their resources and to provide patients with an understanding of where they can seek appropriately skilled care (the ACHD Clinic Directory). Their data (Figure) suggest a nearly doubling in centers self-reporting as providing specialty care to ACHD patients over the past decade, with a concomitant doubling in the number of patient visits to these centers. The importance of these specialized centers is blatantly obvious. Mylotte and colleagues, for instance, demonstrated that ACHD specialty care in Canada was associated with a significant reduction in overall mortality. The American College of Cardiology/American Heart Association ACHD guidelines have recommended that patients with moderate or complex ACHD (=55% of all ACHD patients) be seen at least yearly in such ACHD centers. This means at least 800,000 patient visits to specialized centers yearly. Extrapolating from Adult Congenital Heart Association/International Society of Adult Congenital Heart Disease data and assuming an average of 1.7 visits per patient per year (derived from data from our center), only a paltry 60,000 ACHD patients are currently benefitting from regular specialized care in US ACHD centers (<8% of ACHD patients who require it).

These data emphasize the growing need to train additional physicians skilled in the assessment and management of ACHD. Care of ACHD, despite its complexity and need for expertise, remains not well reimbursed in the United States, and this may detract from its current appeal as a cardiovascular subspecialty. As a result, it will take likely many decades to appropriately expand our ACHD specialist population. Until that time, there is a greater need than ever to disseminate information about ACHD and to instruct general cardiologists and primary practitioners in the community on how to identify these conditions and to recognize when and where to appropriately refer. Too many patients become lost to follow-up once they leave home and the care of their families and local pediatric cardiologists. We must strive to educate and empower these patients to seek continuing medical attention for what they and their families may view as a “cured” condition. With the explosion of social media, the electronic infrastructure is finally ready to get the critical message that CHD requires and benefits from lifelong specialized care to the growing masses of patients with CHD. Gilboa and colleagues have pointed out that the elephant is charging toward us, and it behooves the cardiology community to remove our blinders to the problem and to seriously begin developing the infrastructure to deal with it. We still have a long way to go to meet these needs.

ACKNOWLEDGMENTS
We would like to thank Paula Miller of the American Congenital Heart Association for contributing data from the ACHD Clinic Directory.

DISCLOSURES
Dr Krasuski has been a consultant for Actelion and Bayer Pharmaceuticals and a scientific advisory board member for Ventripoint (unfunded). Dr Bashore reports no conflicts.

AFFILIATIONS
From the Division of Cardiology, Duke University Medical Center, Durham, NC (R.A.K., T.M.B.); and Duke Clinical Research Institute, Durham, NC (R.A.K.).

FOOTNOTES
Circulation is available at http://circ.ahajournals.org.

REFERENCES


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Richard A. Krasuski and Thomas M. Bashore

Circulation. 2016;134:110-113; originally published online July 5, 2016;
doi: 10.1161/CIRCULATIONAHA.116.023370
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/134/2/110

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