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From Numbers to Guidelines

Ariane J. Marelli[a,*], Michelle Gurvitz[b]

[a]McGill Adult Unit for Congenital Heart Disease (MAUDE Unit), McGill University Health Center, Montreal, Quebec, Canada, H3A 1A1
[b]Department of Cardiology/BACH, Children's Hospital Boston, Boston, MA 02115

Abstract Advances in pediatric cardiology and cardiac surgery have resulted in a change in the mortality, prevalence, and age distribution of patients with congenital heart disease (CHD). In this chapter, we review the changing epidemiology of CHD and the impact of these trends on health services utilization and delivery in this population. We demonstrate not only that adult CHD (ACHD) patients have high utilization rates but also that care gaps exist where disease is expected to be lifelong. We outline the components of quality improvement for the care of ACHD patients. We review the newly published guidelines for the management of CHD conditions in adults, anchoring them to structure, process, and outcome indexes of quality of care. We highlight the manpower needs and the importance of an appropriately trained cardiology workforce to provide ACHD care. Finally, we review recommended health care systems ideally suited to deliver care to this population. (Prog Cardiovasc Dis 2011;53:239-246)

Then vs now

Mortality rates from congenital defects have declined over recent decades.[1] From 1979 to 1997, age-adjusted population death rates from all defects declined 39% in the United States from 2.5 to 1.5 per 100,000 or 1.9% per year.[2] During the same observation period, mortality declined for both simple and complex lesions, most notably transposition of the great arteries (TGA), tetralogy of Fallot (TOF), atrioventricular canal defect, and aortic coarctation (AoC).[2] Despite such progress, congenital heart disease (CHD) remains the most common cause of infant death associated with birth defects.[1] Mortality has shifted away from children and toward adults.[3] In a population-based cohort, a total of 8,561 deaths occurred in 71,686 patients with CHD followed for 982,363 patient-years from 1987 to 2005. The proportion of infant and childhood deaths declined over time, exceeding that observed in the general population. Not surprisingly however, long-term survival following surgical repair of complex repair remains compromised, with survival rates decreasing from the third and fourth decade onward.[4]

Although the overall prevalence of CHD has increased over time, population trends indicate proportionally different changes in children and adults. Between 1985 and 2000, the prevalence of severe CHD increased by 85% in adults compared with 22% in children, consistent with the notion that the greatest survival trend has occurred in those with more severe forms of CHD.[5] Prevalence estimates have been generated using birth rates and estimated survival rates for different CHD disease categories.[6-8] In 2000, the number of adult congenital heart disease (ACHD) patients in the United States was estimated to be between 650,000 and 1.3 million.[7,8] Extrapolating the Canadian population data to the United States for the year 2000[5] for a US population of 209 million adults,[9] a minimum of 856,000 patients in the United States were expected to have ACHD in 2000. Using ACHD population growth rates from 1985 to 2000 from the United Kingdom[6] and Canada,[5] 9,000 to 15,000 CHD patients are expected to have graduated to adulthood

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* Address reprint requests to Ariane J. Marelli, MD, MPH, Associate Professor of Medicine, McGill University, Director, McGill Adult Unit for Congenital Heart Disease (MAUDE Unit), McGill University Health Center, 687 Pine Ave West, Room H4-33, Montreal, Quebec, Canada, H3A 1A1.

E-mail addresses: ariane.marelli@mcgill.ca (A.J. Marelli), michelle.gurvitz@cardio.chboston.org (M. Gurvitz).

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Abbreviations and Acronyms

ACC = American College of Cardiology

ACHD = adult congenital heart disease

AHA = American Heart Association

AoC = aortic coarctation

ASD = atrial septal defect

CHD = congenital heart disease

IE = infective endocarditis

HSU = health services utilization

TGA = transposition of the great arteries

TOF = tetralogy of Fallot

each year over the last decade, adding an additional 90,000 to 150,000 new patients to this growing population.

In a Quebec population-based study, the prevalence of CHD in the year 2000 was 4/1,000 adults and 12/1,000 children in the year 2000.[5] There was a predominance of women relative to men in adults with severe forms of CHD; this is also true relative to the normal population.[5] In the year 2000, prevalence rates of CHD were higher in children than adults, with CHD present in 1/84 children and 1/245 adults.[5] Because the ratio of adults to children in Canada and the United States is approximately 1 child for every 3 adults,[9,10] however, the overall number of adults exceeds the number of children with CHD; and the number of adults and children with severe CHD was nearly equal by the year 2000.[5] As a result of observed demographic changes over the last 2 decades, the overall CHD population has aged, most notably in those with severe forms of CHD, where the mean age increased from 11 years old in 1985 to 17 years old in 2000.[5] The median age of the entire ACHD population was 40 years old in 2000 and 29 years old in the subset of adults with severe CHD.[5]

Thus, overall mortality of children born with CHD has declined, resulting in a larger number of CHD patients reaching adulthood, including those with severe CHD who previously did not survive. This increases the overall age of the population and results in more adults alive with CHD than children, with a predominance of adult women. These demographic changes shift the burden of lifelong disease care into the adult population and are expected to influence the distribution of health service utilization in the CHD population.

Impact on health services utilization

There are limited data documenting frequency of health services utilization (HSU) in the ACHD population. From 1996 to 2000, overall outpatient and inpatient service utilization in 22,096 ACHD patients in Quebec resulted in outpatient physician visits of a median of 6 times per year among 90% of the population.[11] Utilization of inpatient services on a population level revealed that, over a 5-year period, 50% of ACHD patients were hospitalized on a medical or surgical service and 16% required critical care.[11] Among 6 large regional ACHD centers worldwide, an average of 10% of patients were hospitalized each year.[12] At the Royal Brompton, the fastest-growing segment of users of outpatient health services in ACHD patients was 20 to 40 years of age, whereas those older than 30 years constituted the most rapidly growing segment of hospitalized patients.[6] Examination of utilization of emergency department services and HSU during the transition years is particularly revealing. Population data on 22,096 ACHD patients showed that 68% visited the emergency department at least once over the 5-year period.[11] Hospitalization patterns during the transition between adolescence and adulthood in 9,017 hospitalizations in the state of California from 2000 to 2003 showed that although overall hospitalization rates decreased in patients 17 to 23 years old, a higher proportion of admissions occurred via the emergency department.[13] In 643 Canadian patients diagnosed with CHD before the age of 6 years and followed to age 22 years, only 39% of the cohort saw a cardiologist between 18 and 22 years of age.[14] Even among those with severe lesions, only 79% were seen by a cardiologist in early adulthood.[14] These data reveal that HSU rates in this population are high and that a particularly disturbing trend in dispersion of care occurs during the transition period. Taken together, these findings suggest that needs for health services, especially in the transfer of care from pediatric to adult health care system, are not being met and that quality of care stands to be improved. Further to the point, these needs will only increase as the population ages.

Improving quality of care

The purpose of improving quality of care for ACHD patients is to provide health services that increase the likelihood of desired health outcomes and are consistent with current professional knowledge.[15] The goal is to decrease variations in practice patterns; avoid care gaps in patients with lifelong conditions; and provide a currency for dialogue among patients, health care providers, the health care system, and policy makers. Quality of care can be evaluated in terms of structure, process, and outcome measures.[16] Structural parameters relate characteristics of physicians and hospitals or health delivery systems. These include recommendations that emphasize consultation with an ACHD specialist or ACHD centers. Process parameters relate to components of the health care encounter. These include lesion-specific recommendations for frequency of follow-up and type of investigations. Outcome parameters are those expected to relate to the patient's subsequent health status. These include medical, percutaneous, or surgical interventions that are expected to impact the patient's morbidity or mortality. One method of