Pediatric Cardiac Surgery
The Long View

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Congenital heart defects are the most common birth defects. Many of them are associated with substantial morbidity and mortality. Of all congenital conditions, heart defects account for the highest resource utilization among pediatric hospitalizations in developed countries. The history of cardiac surgery now spans nearly 8 decades. In the earliest era of cardiac surgery, operations on children and young adults with congenital cardiac malformations accounted for the lion’s share of “heart operations.” Closure of patent ductus by Strieder and by Gross in Boston, MA (1937, 1938), repair of coarctation of the aorta by Craaford in Stockholm, Sweden (1944), and the “blue baby” shunt procedure by Blalock in Baltimore, MD (1944) were landmark achievements that signaled the birth of the medical and surgical subspecialties of pediatric cardiology and congenital heart surgery. What followed was a period of 7 decades marked by huge increases in the understanding of circulatory physiology and the abnormal morphology and functional consequences of structural congenital heart disease; burgeoning technology that may facilitate earlier, more precise, and more complete characterization of abnormally formed hearts; and therapeutic innovations that may save and prolong lives and ultimately contribute to the mission of maximizing longevity and optimizing the quality of life of individuals living with congenital heart disease. By the 1970s, primary repair of complex forms of congenital heart disease in neonates and young infants was becoming a more widely acknowledged reality. By the mid-1980s, virtually all common congenital cardiac anomalies had been moved to the “treatable” column on the tables expressing severity. The rapid progress in surgical therapy was just one element of a bigger picture that also encompassed the application of less invasive and more informative diagnostic modalities, the emergence of pediatric cardiac critical care, and the evolution of a true multidisciplinary approach to management of congenital heart disease, increasingly informed by advances in cell biology, pharmacology, and other basic sciences.

By the beginning of the 21st century, contemporary estimates suggested that 85% of patients born with congenital heart disease in the United States would survive into adult life. By 2010, researchers reported that approximately 2 million infants, children, adolescents, and adults were living with congenital heart disease in the United States and that for the first time, the number of adults in the United States living with congenital heart disease was as large as the number of children living with congenital heart disease. Investigators in other developed countries produced estimates that paralleled the trends and inferences drawn from US data. Cross-sectional regional population-based studies have yielded valuable information concerning the epidemiology of congenital heart disease. Multicenter national and international surgical databases provide detailed and accurate accounts of risk-adjusted surgical outcomes. Around the world, efforts to collect data for quality assurance and for benchmarking have led to the recognition that data sets that extend over many decades with reliable follow-up and with complete life status information are extremely important to our growing understanding of the progress that has been made and of the challenges that lie ahead, both in terms of the effectiveness of disease-specific and patient-specific approaches to diagnosis and management and in terms of the healthcare and societal resources that will be required to support continued progress and a growing population of patients surviving and living with congenital heart disease.

This issue of Circulation contains 2 very significant reports in which investigators from Finland and Norway present data pertaining to cardiac operations performed on children over periods of 4 and 6 decades, respectively. Raissadati and associates from Helsinki report data pertaining to all patients who underwent pediatric heart surgery during childhood in Finland from 1953 to 2009, and Erikssen and associates from Oslo report data pertaining to nearly all patients who underwent heart surgery during childhood in Norway from 1971 to 2011 (including all patients from 2003–2011). What is most remarkable about these 2 reports is that both groups of investigators were able to use national population registries to obtain recent life status data for >98% of patients. In all of Finland, 10,964 patients ≤15 years of age underwent 13,786 heart surgery operations from 1953 to 2009. In Oslo, 7,038 patients aged ≥16 years underwent 9,380 heart surgery operations in 1971 to 2011 (accounting for ≈80% of operations in all of Norway before 2003 and 100% of operations since 2003). Some “back of the napkin” calculations, assuming an average population of approximately 5 million in Finland and approximately 4 million in Norway over the period of interest, yield an average of 246 pediatric heart operations per year in Finland (roughly 0.05
per 1000 population) and 234 pediatric heart operations per year in Norway (roughly 0.06 per 1000 population). In general, the number of operations per year increased from one era to the next. This is explained mostly by the fact that the number of patients undergoing operations for complex defects increased progressively across eras. The trends in numbers of operations for more simple defects were less consistent, potentially related to the emergence and availability of alternative nonsurgical therapies.

Several important but not surprising findings were shared by both groups of investigators. In each study, the average age at first operation decreased markedly over the study period. Although slightly different schemes were used to stratify operations according to relative complexity, each longitudinal series was characterized by an increasing fraction of all operations being performed for more complex defects over the duration of the study interval. The early mortality decreased significantly across years for virtually all diagnostic groups of intermediate and high complexity (the mortality for very low complexity categories having been relatively low to begin with). In the aggregate, the 2 studies revealed that long-term survival of patients who underwent surgery in a more recent era was significantly better than it was for those operated on in an earlier era, although changes in very long-term survival (eg, 40–60 years) were often influenced by small numbers of deaths, and not all late deaths were of cardiac origin. Both studies included analyses of survival within individual strata of complexity and within representative diagnostic groups. In both study cohorts, the more recent rise in the fraction of patients with more complex or more “severe” defects was accompanied by a rise in the average number of operations per patient over the course of the entire study period. However, the investigators in Oslo observed that there was a significant increase in reoperation-free survival among patients who had surgery in 2000 to 2011 compared with those from 1990 to 1999. An additional common theme is the historical pattern by which cardiac operations on children were concentrated into a smaller number of hospitals and eventually fully centralized. In Finland, pediatric cardiac operations had been performed at 5 university hospitals and 1 district hospital. Since 1997, all pediatric cardiac operations in Finland have been performed at Helsinki University’s children’s hospital. In Norway, before 2003, ≈80% of all pediatric cardiac surgeries had been performed at Oslo University Hospital, Rikshospitalet. Virtually all pediatric cardiac operations in Norway have been performed there since 2003.

So, these 2 groups of investigators from Helsinki and Oslo, who had the foresight to establish comprehensive cardiac surgical databases many decades ago and to maintain and curate them to ensure their usefulness, provide information of a nature that can be obtained from only a few sources. The picture is significantly enhanced by contemporaneous life status analysis, for which national mortality statistics provide very nearly complete information. The general impression that one gleaned from these 2 reports includes many ideas that we may have taken for granted previously, although in some respects, we may have done so without sufficient proof. These include the following:

1. The idea that in developed countries with reasonably universal access to tertiary healthcare services for all patients at all ages, the number of patients undergoing heart surgery as neonates, infants, and children is going up and is not accounted for entirely by changes in birth rates.

2. The observation that the age at first surgery has gone down rather dramatically over several decades, and this is primarily related to the fact that patients with increasingly complex forms of heart disease can now be managed surgically very early in life, before they succumb to the consequences of impaired circulation. This, of course, extends to hypoplastic left heart syndrome and related anomalies that share the characteristics of functionally univentricular physiology and ducal dependency of the systemic circulation. Before the mid-1980s, these malformations were widely considered to be “uniformly lethal.”

3. The reality that both short-term and long-term outcomes, with respect to survival, have improved substantially and continue to improve for the most challenging forms of congenital heart disease. Among these are anomalies such as transposition of the great arteries, for which “anatomic repair” offers a very good chance of restoring normal or near-normal cardiovascular physiology. Also among these are anomalies such as tetralogy of Fallot, for which the expectation of survival after initial reparative surgery is extremely high but for which subsequent interventions to preserve the function of the right ventricle will be a reality for many, if not most, patients.

4. There are anomalies such as interrupted aortic arch with ventricular septal defect and left ventricular outflow obstruction, for which survival after neonatal surgery that preserves biventricular physiology has improved greatly over earlier decades but which carries a substantial risk of the need for subsequent interventions.

Finally, as much as we are assured by the encouraging findings in these important reports from Finland and Norway, it is impossible to escape the realization that even this kind of population-based data gives us just a small glimpse of a piece of the big picture that describes management of congenital heart disease. Surgery is but one important part of a
multidisciplinary approach. In addition, in the current era, across-the-board operative mortality for all patients in large international, multi-institutional congenital heart surgery registry databases is on the order of 4%, or slightly less. If our focus is primarily on in-hospital mortality, then we are overlooking the 96% of patients who survive to hospital discharge and failing to consider the important morbidities that they may experience. Unless we pursue a better understanding of those morbidities, we cannot have a rational, evidence-based approach to extending survival, mitigating morbidities, and helping to optimize the functional status and quality of life of patients with congenital heart disease.

The emergence of an ever-larger population of patients surviving to adulthood with congenital heart disease reflects very positively on the community of individuals and organizations that have cared for these patients over the past many decades. But it is a clarion call announcing the reality that with the possible exception of the simplest anomalies, the vast majority of forms of congenital heart disease are actually chronic diseases with lifelong implications for patients’ health and well-being and for the societal platforms and healthcare systems that must adapt to ensure that congenital heart disease patients of all ages have access to appropriate professional expertise and care. As for the data that we collect and analyze, we must learn to move beyond the status quo, wherein a patient’s surgical history is sequestered in one silo, their history of diagnostic data and interventions in another, and information relevant to their functional status and even their genotype in others still. And, as the investigators in Helsinki and Oslo have illustrated, databases must be truly longitudinal. This is not a time to abandon cardiac surgical databases as being too limited in scope, nor a time to call for a reinvention of the wheel. Too pliant unique patient, surgeon, and hospital identifier fields in the STS database. Achievements in congenital heart defect surgery: a prospective, 40-year study of 7038 patients. Circulation. 2015;131:337–346.


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