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, they account for the highest resource utilization among pediatric hospitalizations in developed countries.\(^1\) The history of cardiac surgery now spans nearly eight 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 (1937, 1938), repair of coarctation of the aorta by Craaford in Stockholm (1944), and the “blue baby” shunt procedure by Blalock in Baltimore (1944) were landmark achievements that signaled the birth of the medical and surgical sub-specialties of pediatric cardiology and congenital heart surgery. What followed was a period of seven 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, which 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 multi-disciplinary approach to management of congenital heart disease, increasingly informed by advances in cell biology, pharmacology, and other basic sciences.
By the beginning of the twenty-first century, contemporary estimates suggested that 85% of patients born with congenital heart disease in the U.S. survive into adult life.\(^2\) By 2010, researchers reported that about 2 million infants, children, adolescents, and adults were living with congenital heart disease in the United States, and estimated that for the first time, the number of adults in the U.S. living with congenital heart disease was as large as the number of children living with congenital heart disease.\(^3\) Investigators in other developed countries produced estimates that paralleled the trends and inferences drawn from U.S. data.\(^4\) Cross-sectional regional population based studies have yielded valuable information concerning the epidemiology of congenital heart disease.\(^5\) Multi-center national and international surgical databases provide detailed and accurate accounts of risk-adjusted surgical outcomes.\(^6\) Around the world, efforts to collect data for quality assurance and for benchmarking has led to the recognition that datasets 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 also 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 two very significant reports in which investigators from Finland and Norway present data pertaining to cardiac operations performed on children over periods of four decades and six decades, respectively. Raissadati and associates\(^7\) from Helsinki report data pertaining to all patients who underwent pediatric heart surgery during childhood in Finland from 1953-2009, and Erikssen and associates\(^8\) from Oslo report data
pertaining to nearly all patients who underwent heart surgery during childhood in Norway from 1971-2011 (including all patients from 2003-2011). What is most remarkable about these two reports is that both groups of investigators were able to utilize national population registries to obtain recent life status data for greater than 98% of patients. In all of Finland, 10,964 patients, age 15 years or less underwent 13,786 heart surgery operations from 1953-2009. In Oslo, 7038 patients age 16 years or less underwent 9380 heart surgery operations in 1971-2011 (accounting for approximately 80% of operations in all of Norway prior to 2003, and 100% of operations since 2003). Some “back of the napkin” calculations, assuming an average population in Finland of about 5 million over the period of interest, and in Norway of about 4 million, yield an average of 246 pediatric heart operations per year in Finland (roughly .05 per 1000 population) and 234 pediatric heart operations per year in Norway (roughly .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 non-surgical therapies.

Several important, but not surprising findings were shared in common by both groups of investigators. In each study, the average age at first operation decreased markedly over the study period. While slightly different schemes were employed 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 two studies revealed that long-term survival of patients that underwent surgery in a more recent era was significantly better than it was for those operated in an earlier era, though changes in very long-term survival (e.g. 40-60 years) were often influenced by small numbers of deaths, and not all late deaths were of cardiac origin. Both studies include 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 2000-2011, when compared to those from 1990-1999. 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 five university hospitals and one district hospital. Since 1997 all pediatric cardiac operations in Finland have been performed at Helsinki University’s children’s hospital. In Norway, prior to 2003, about 80% of all pediatric cardiac surgery had been carried out at Oslo University Hospital, Rikshospitalet. Virtually all pediatric cardiac operations in Norway have been carried out there since 2003.

So, these two 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 insure their usefulness, provide information of a nature that can be obtained from only a small number of 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 glean's from these two reports includes many ideas that we may
previously have taken for granted, though in some respects we may have done so without sufficient proof. These include:

1- The idea that in developed countries with reasonably universal access to tertiary health care 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 sharing the characteristics of functionally univentricular physiology and ductal 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 “anatomical 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 following initial reparative surgery is extremely high, but for which the potential need for subsequent interventions to preserve the function of the right ventricle will be a reality for many, if not most patients. Then, there are anomalies such as interrupted aortic arch with ventricular septal defect and left ventricular outflow obstruction, for which survival following neonatal surgery that preserves biventricular physiology has improved greatly over
earlier decades, but which carries substantial risk of the need for subsequent interventions.\textsuperscript{11} And finally, there are the anomalies where univentricular physiology is the best that can be achieved without heart replacement. A staged reconstructive approach, carried out over the first several years of life, has produced results that are dramatically better than those achieved a few decades ago. But considerable uncertainty remains concerning factors that influence the durability of the functionally univentricular “Fontan circulation.”\textsuperscript{12}

4- The idea that improved survival has been accompanied by a pattern of increasing interventions during a patient’s lifetime. Though only surgical interventions are comprehensively considered in these two studies, it is nonetheless apparent that improving long-term survival may involve not only the possibility of an individual patient having multiple surgical procedures, but also catheter-directed interventions including relief of obstructions at the levels of vessels and valves, device implantations, and interventions to address arrhythmias.

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 multi-disciplinary 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 four percent, 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.\textsuperscript{13} And 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 last many decades. But it is a clarion call announcing the reality that, with the possible exception of the very 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 health care systems that must adapt, in order 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 much of value has been accomplished, as evidenced by these two reports. Rather it is a time to leverage the wealth of information that is sequestered in individual sub-specialty databases, by making use of proven, as well as newer and potentially better innovative approaches to linking datasets.\(^\text{14}\) It is a time to explore the utility of universal unique identifiers that protect the confidentiality of individuals’ health information,\(^\text{15}\) and a time to make the information in a nation’s death master file available to appropriately credentialed investigators in universities and professional societies, as it currently is in Finland and Norway.\(^\text{16}\)

**Conflict of Interest Disclosures:** None.
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