Late Causes of Death After Congenital Heart Defects
A Population-Based Study From Finland*

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“Life can only be understood backwards; but it must be lived forwards.”
—Søren Kierkegaard (1)

Improvement in life expectancy of patients with congenital heart disease (CHD) (2,3) has resulted in emergence of a patient population for which there is sparse track record in adult medicine. Tracking these outcomes is fascinating, yet problematic because CHD is not a single disease entity but an infinite combination of structural and physiological variables. As medical and surgical management has evolved, so has the patient population, due to a continually shifting selection bias. In earlier times, prostaglandin was not available to stabilize newborns with ductal-dependent lesions. Patients with chromosomal abnormalities were often excluded from surgical repair. Fetal diagnosis and neonatal transport became available in more recent times. Therefore, survivors of an earlier era represent a hardier and less complex group whereas more recent times have seen survival of a more marginally viable population.

In this issue of the Journal, Raissadati et al. (4) followed mortality after surgery for CHD for up to 60 years. This study is unique because of a remarkable 97.5% follow-up of longitudinal follow up and precisely matched normal controls provided by Finnish registries. These datasets have an advantage over hospital or insurance data, which only accounts for patients with access to care and is subject to coding variability (5). Moreover, reliable longitudinal data cannot be tracked within a system in which patients move between insurers and institutions.

The study (4) includes CHD patients operated on in Finland between 1953 and 2009, divided into early (1953 to 1989) and late (1990 to 2009) eras, loosely coinciding with consolidation of complex surgery to 1 institution in the mid-1990s. Patients were grouped into simple (acyanotic), complex (cyanotic), and miscellaneous lesions. Causes of death attributed to CHD were classified as congestive heart failure, sudden, and perioperative, with the remainder classified as cardiovascular. Although there is considerable overlap and ambiguity among these categories, the approach is practical and easily duplicated.

For patients with simple defects, the dramatic decrease over time in early post-operative mortality and deaths from cardiovascular causes is likely due to earlier intervention and initial complete repair, resulting in a lowered incidence of pulmonary vascular disease and ventricular failure. In the earlier era, 86% of post-operative deaths after ventricular septal defect repair were among patients with previous palliation. For entire group of simple lesions, 40-year survival was 89% among initial post-operative survivors, with the majority of deaths due to noncardiac causes.

For complex lesions, advances over the years were more modest, the majority of deaths occurred within 10 years and initial post-operative survivors had a 40-year survival of 65%. Post-operative tetralogy of Fallot mortality declined in the latter era, probably due to early, 1-stage repair. Mortality also decreased in

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patients with transposition of the great arteries during the later era, which coincides with a change from the atrial to the arterial switch approach. Late mortality remained high for patients with single-ventricle patients, however.

A bimodal distribution of mortality from congestive heart failure was found for most lesions, with higher mortality in the first few years of life, followed by a relatively benign course lasting more than 20 years for most simple lesions and <10 years for complex patients. This is consistent with resource utilization data from the United States that reflects a relatively steady period after the infancy, followed by increased hospitalization as they approach emerging adulthood (6,7). Raissadati et al. (4) postulate that the rising mortality could be related to transfer of care to an adult health system, but one might also argue that this pattern reflects the natural history of CHD and that patient transfer during a period of stability is well timed.

Among the non-CHD causes of death, respiratory disease was the most common with more than one-half occurring among patients with a mental disability. Deaths due to neurological (stroke, seizures) and infectious diseases were also more prevalent than the general population. The lack of progress in mortality reduction in these categories is likely due to the superimposition of co-morbid or acquired conditions (8–10). The importance of age-related acquired complications in the adult with CHD reinforces the importance of their care being delivered in an environment that includes the full spectrum of expertise in the recognition and treatment of both congenital and acquired diseases.

A higher incidence of cancer deaths in CHD patients reinforces previously reported concerns (11,12). In the more recent era, the relative risk of cancer in women was 5.90 compared with the general population. While technical advances continue to reduce radiation at each exposure, there is ongoing innovation in the use of invasive procedures and radiologic imaging that adds significant clinical value to patient outcomes, thus the need for continued innovation in echocardiography and magnetic resonance imaging as a substitute for radiologic imaging, particularly for patients with complex lesions.

Deaths due to alcohol use and accidents were higher in the patient population, with the average age of death from accidents at 29 years and a significantly higher incidence in females operated upon in the earlier era. One wonders whether these findings could be related to a higher incidence of mood disorders or associated neurodevelopmental abnormalities, such as visual-spatial coordination or executive functioning and suggests a role for more widely applied screening and preventive measures.

In summary, a comprehensive national registry provides important and necessary insights into long term outcomes of CHD. While we take pride in the substantial improvement in life expectancy that has resulted from more aggressive and centralized surgical intervention, we are also challenged to further maximize the opportunities for a long and productive life. Some problems are related to superimposition of acquired diseases caused by unhealthy aging and others are triggered by the very procedures that permitted survival in the first place. Further advances will require meaningful longitudinal observations from registries like that in Finland. We should focus more attention on early life-style modification to prevent superimposed, acquired diseases. The evidence for increased cancer risk, particularly in young women, should spur more investigation into acceptably safe levels of radiation exposure and the development of evidence-based guidelines for cancer surveillance in the CHD population. The importance of acquired disease prevention in addition to present disease management must be impressed on families and patients at the earliest possible age. To paraphrase James Burke in the long-ago BBC series, Connections: Inventions trigger changes in totally unrelated fields for which the only remedy is continued invention. For that reason, we may never reach the “Finnish-line.”

**REFERENCES**


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