

Causes of Late Mortality of Children Surviving Initial Congenital Heart Surgery: A Report from the Pediatric Cardiac Care Consortium

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Background: Children living with operated congenital heart disease (CHD) continue to be at risk for causes of death that are different from the general population. We used the Pediatric Cardiac Care Consortium (PCCC), a large US-based registry for interventions for CHD, to examine the underlying causes of death (COD) of children operated at <21y of age for a CHD.

Methods: This is a retrospective cohort study of children who were operated for CHD at 45 centers in the PCCC between 1982 and 2003 and were linked with the US-National Death Index (NDI) through 2014. Data provided by NDI-Plus included International Classification of Disease codes for underlying COD and associated morbidities/events contributing to death. Cause-specific mortality for individuals with operated CHD was compared to the general population with adjustment for age and sex. Patients with known chromosomal abnormalities or inadequate identifiers were excluded from the analysis.

Results: Among 31,132 patients surviving their initial congenital heart surgery there were 2,527 deaths (8.1%) over a follow-up period of 33 years (median 18.1, IQR:14.5 -22.2). Median age at death was 1.8 year (IQR: 0.5-12.8). COD varied by severity and type of CHD but, overall, 83.2% of late mortality was associated with CHD or another cardiovascular condition (Figure 1A). As patients aged, the proportion of deaths caused by external causes and neoplasms increased (Figure 1B). The overall standardized mortality (SMR) for patients with operated CHD compared to the general population was 22.6 (95% CI: 19.5-25.7) from external causes and 5.8 (95% CI: 4.2-7.3) from neoplasms.

Conclusion: Patients operated for even the mildest forms of CHD are at high risk for premature cardiovascular death directly or indirectly associated to their underlying CHD warranting continued monitoring to address associated morbidities.

