Decreasing mortality in children with severe congenital heart defects (CHD), 1994-2009

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Objectives: Despite improved diagnostic tools and new therapeutic options, congenital heart defects (CHD) are still an important cause of death in young age. In this nationwide cohort study we describe time trends in two-year mortality among all children with severe CHD born in Norway 1994-2009.

Methods: Information on children with CHD was ascertained from national health registers (The Medical Birth Registry of Norway and the Norwegian Cause of Death Registry) and databases (Oslo University Hospital's clinical database for congenital heart defects and the Cardiovascular Disease in Norway (CVDNOR)) by use of specific diagnostic codes. All children were followed until 31 December 2012. All individuals with CHD codes were assigned to only one diagnostic group by a hierarchical system and classified as having severe (heterotaxy, conotruncal defects, atrioventricular septal defects, anomalous pulmonary venous return, left ventricular outflow tract obstruction, right ventricular outflow tract obstruction (except pulmonary valve stenosis) and other complex defects) or non-severe CHD (septal defects, isolated patent ductus arteriosus, other specified CHD, pulmonary valve stenosis and unspecified CHD). Cumulative mortality proportion (CMP) was defined as the proportion of deaths regardless of cause in a specified cohort during a given period. Time trends were analyzed using Joinpoint Regression Program and are presented as the expected annual percent changes with 95% confidence interval (CI), describing trends by periods, using the best-fit model.

Results: Among the 943,871 live births in Norway 1994-2009, CHD were identified in 11,272 (1.2%) children. The proportion classified as having severe CHD was 23.7% (n=2,673). The two-year cumulative mortality proportion (CMP\textsubscript{2year}) during the entire study period was 17.4% (n=466) in children with severe CHD and 0.3% (n=3,044) in children without CHD. The CMP\textsubscript{2year} per year of birth in children with severe CHD decreased significantly during the study period with an annual percent change (APC) of -3.7% (95% CI: -5.9, -1.4) (figure).

Conclusions: In Norway, the two-year mortality among children with severe CHD has decreased during the period 1994-2009, but is still high compared to the general population. Further investigations are needed to clarify the factors of importance for death in children with severe CHD.