What are the causes of death in patients with congenital heart defects? An analysis of data from the German National Register for Congenital Heart Defects.

Competence Network for Congenital Heart Defects, Berlin, Germany (1); National Register for Congenital Heart Defects, Berlin, Germany (2); Saarland University Medical Center, Department of Paediatric Cardiology, Homburg, Germany (3); Children’s Heart Centre Sankt Augustin, Department of Paediatric Cardiothoracic Surgery, Sankt Augustin, Germany (4); Münster University Hospital, Center for Adults with Congenital Heart Defects, Münster, Germany (5); Heart and Diabetes Center NRW, Center for Congenital Heart Defects, Bad Oeynhausen, Germany (6); Hannover Medical School, Centre for Internal Medicine>Department of Cardiology and Angiology, Hannover, Germany (7)

Introduction:
Thanks to progress in paediatric cardiology, cardiology, cardiac surgery and intensive care, the mortality rates of patients with congenital heart disease (CHD) was reduced significantly in the past decades. However, the mortality rates in CHD patients are still higher than in the general population. The aim of further improvements calls for a detailed analysis of the causes of death. So far, such an analysis has not been available in Germany.

Methods:
The German National Register for Congenital Heart Defects (NRCHD) was scanned systematically for deceased patients. Deceased patients with a confirmed diagnosis of CHD were included. Data relating to the cardiac diagnosis, symptoms, operations, interventions, concomitant diseases and causes of death were used for analysis. Two age groups were formed (< 18 years; ≥ 18 years).

Results:
133 deceased patients were included in the analysis (median age at the time of death: 20.6 ± 19.4 years; 42.1% female). Of those, 12.8% had mild, 28.6% had medium and 58.6% had complex CHD. In 117 patients, the CHD was the direct cause of death (88%), 67 (57%) of those died perioperatively. 16 patients (12%) died from non-cardiac causes. 75 patients (56.4%) died before their 18th year of life, 58 patients (43.6%) afterwards. The two age groups did not differ with respect to sex distribution (p = 0.36), complexity of CHD (p = 0.36), NYHA status (p = 0.27), presence of cyanosis (p = 0.29) or cause of death (died from the CHD: p = 0.06). There was also no difference regarding the frequency of perioperative death (p = 0.56). Regarding concomitant diseases, pulmonary diseases (p = 0.02) and renal diseases (p = 0.008) were found significantly more often in patients over 18 years.

Conclusions:
The analysis of causes of death in patients with CHD enrolled in the NRCHD revealed that the deceased patients frequently had complex CHD and that the majority of them died as a consequence of their CHD. The fact that approximately two thirds of named deaths occurred perioperatively highlights the need to investigate strategies to optimise both the time of re-operation and operation methods including catheter based alternatives.