Long-term survival in patients with univentricular heart: a nationwide, register-based cohort study

A.G. Ozturk1, M. Dellborg2, K.W. Giang2, M. Dalen3, P. Sorensson1, Z. Mandalenakis2

1Karolinska Institute, Department of Medicine Solna, Stockholm, Sweden; 2Institute of Medicine - Sahlgrenska Academy - University of Gothenburg, Gothenburg, Sweden; 3Karolinska Institute, Department of Molecular Medicine and Surgery, Stockholm, Sweden

Funding Acknowledgement: Type of funding sources: Public grant(s) – National budget only. Main funding source(s): Swedish heart lung foundation

Background: Univentricular heart (UVH) is a broad term that covers various cardiac structural abnormalities in which one ventricle is severely underdeveloped, or a ventricular septal wall did not form, leading to a functional single-ventricle physiology. Patients with UVH have a very limited life expectancy if not surgically treated. While the outcome in children with UVH have been extensively studied, data regarding the long-term outcomes in an unselected, nationwide cohort is still lacking.

Purpose: The aim of the present study was to determine long-term survival in patients with UVH compared to an age- and sex-matched control population without congenital heart disease in Sweden.

Methods: We linked data from the Swedish Patient and Cause of Death Register to identify patients with UVH that were born between 1970 and 2017. Each patient with UVH was matched with ten individuals without congenital heart disease for year of birth and sex. The mortality risk in patients with UVH compared with matched controls was analyzed using Cox proportional regression models and Kaplan-Meier survival analysis.

Results: Of a total of 5,075 patients with UVH and 50,620 matched controls, 1,992 (37.9%) and 485 (1.0%) died, respectively. The mean follow-up (standard deviation) was 15.7 (± 14.2) years. A total of 758 patients (14.9%) of the UVH population were diagnosed with hypoplastic left heart syndrome (HLHS). The mortality risk was 53.01 (95% CI, 48.0–58.6) times higher in UVH and 163.5 (95% CI, 124.3–215.2) times higher in HLHS compared to controls. According to birth period, highest mortality was found at the earliest UVH cohort (1970–1981) 59.8%, and decreased by later birth periods. The lowest mortality was in the latest birth period, 2006–2017 (16.3%).

Conclusions: In this nationwide, register-based cohort study, the overall risk of mortality was more than 50 times higher in patients with UVH compared to matched controls. However, an increased survival was observed over time and by later birth periods. Quadragenarians with UVH is still a rare group but are expected to increase during the next decades.