Objective. To collect reliable and complete data for evaluation of long term survival after surgery for univentricular heart defects. 

Methods. All 304 patients (126 girls/178 boys) operated due to a univentricular heart defect in our institution before the age of 18 years from Jan 1st 1994 to Jan 1st 2009 were included. Patient files were cross-checked as of Jan 1st 2012 against the National Population Registry in Sweden allowing for reliable and complete data on survival. Two patients (0.6%) emigrated and were lost to follow up. 

Results. Median age and weight at first surgery was 10 days (0-15.1 years) and 3.6 kg (1.2 – 30.0). Median age of survivors at follow up was 11.4 years (3.0 – 30.7). 83 (27.3%) deaths occurred with a median age at death of 69 days (0.01-22.6 years). Median survival time in the deceased patients was 29 days (0- 11.2 years) after the last major surgery. Mortality in 271 patients with their first surgery in 1994-2008 were 51/110 (46.3%) in patients with classic hypoplastic left heart syndrome or an unbalanced atrioventricular septal defect with left ventricular hypoplasia (A), compared to 9/87 (10.3%) in patients with tricuspid atresia, double inlet left ventricle or pulmonary atresia with intact ventricular septum (B) (p< 0.001). Mortality in children with an indeterminate ventricular morphology and/or other complex congenital heart defects (C) was 18/74 (24.3%). Mortality in patients with their first surgery in 1994-1998, 1999-2003, and 2004-2008 was 41/95 (43.1 %), 19/87 (21.8%) and 18/89 (20.2%) respectively (p< 0.05). Such an improvement was found in all three subgroups and in patients with their first surgery in 2004-2008 mortality was 13/33 (39%), 2/32 (6%) and 3/24 (12.5%) in group A, B and C respectively. Heart transplantation was performed in 16 patients (5%) with four late deaths. 

Conclusion. Long term survival in patients with a systemic ventricle of left ventricular morphology was high (89.7%), while survival in patients with a systemic ventricle of right or indeterminate ventricular morphology was worse, i.e 53.7% and 75.7% respectively. An improvement of survival was seen over time in all groups.