Causes of late death after paediatric cardiac surgery in patients with univentricular heart defects

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Objectives
To identify those patient groups at risk for late mortality after surgery for univentricular heart defects by assessing the mode of their late deaths.

Methods
All 306 patients operated for univentricular heart defects at our institution before the age of 18 years between Jan 1st 1994 and Jan 1st 2009 were cross-checked against the Swedish National Population Registry on January 1st 2012 to reliably identify all dead patients. Of 83/306 deceased patients 41/306 (13.4%) had died more than 30 days after the last surgery and 19/306 (6.2%) had undergone a heart transplantation. 2/306 patients (0.7%) were lost to follow-up (emigration). The circumstances of late death were analysed by reviewing clinical charts and autopsy reports. The mode of death was defined as the condition that initiated a clinical course of deterioration leading to death rather than the pre-terminal event causing the patient to pass a terminal threshold in an already marginal physiological state.

Results
The mode of death was considered to be related to the underlying heart defect in 34 and most likely CHD-related in further 5 cases together accounting for 39/41 (95%) of all late deaths. Of the late deceased patients 29/41 (71%) had a systemic ventricle of right ventricular morphology. 23/41 (56%) patients died after the first palliation mainly due to shunt complications (7/23 – 30%), circulatory failure in the face of infection or pulmonary disease (6/23 – 26%) and acute, autopsy-negative circulatory collapse in an ambulatory setting (5/23 – 22%). Other causes included ventricular failure and myocardial ischemia. The main mode of death after the Glenn operation was ventricular failure (5/10) and after the Fontan operation thromboembolic complications in 2/4. 4/41 patients died due to complications after heart transplantation (3 malignancies, 1 rejection). Syndromes and/or relevant congenital defects were noted in 4/41 (9.8%).

Conclusion
Deaths in patients with univentricular hearts were nearly exclusively related to their heart defect and occurred predominantly after the first palliation. The main mode of death was a failing shunt circulation. The majority of patients had a systemic ventricle of right ventricular morphology. Associated syndromes and/or congenital defects were relatively uncommon.