Causes of late death in patients with congenital heart defects after paediatric cardiac surgery aiming at biventricular repair

Polte C., Berggren H., Sunnegårdh J.  
Departments of Paediatric Cardiology and Paediatric Surgery, The Queen Silvia Children’s Hospital, Sahlgrenska University Hospital, Gothenburg, Sweden

Objectives
The aim of the study was to identify those patient groups at risk for late mortality after surgery for congenital heart defects (CHD) aiming at biventricular repair by assessing the mode of their late deaths.

Methods
All 2132 patients operated for CHD aiming at biventricular repair 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 all 101/2132 (4.7%) deceased patients 70/2132 (3.3%) had died more than 30 days after the last surgery and 6/2132 (0.3%) had undergone a heart transplantation. 21/2132 patients (1.0%) 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.

Results
The mode of death was considered to be CHD-related in 42 and most likely CHD-related in further 9 cases together accounting for 51/70 (73%) of late deaths. The 9 deaths without a definite cause occurred in patients with complex heart defects and in 7/9 patients suddenly in an ambulatory setting. In the majority of these 9 cases an arrhythmia is highly probable. The dominating mode of CHD-related deaths was ventricular failure in 20/42 (48%), which was in 11/20 (55%) due to pulmonary hypertension of different aetiologies including pulmonary venous stenosis (5) and AVSD or VSD in Down syndrome (3). 8/42 (19%) died because of a failing shunt-circulation (shunt-occlusion 2, circulatory failure 6). 19/70 (27%) of all late deaths were not related to the underlying congenital heart defect. Of all patients 49/70 (70%) had a syndrome, chromosomal and/or congenital defects.

Conclusion
Late mortality after surgery for CHD aiming at biventricular repair is low and in nearly one third of cases the deaths were not related to the underlying cardiac defects. Syndromes and relevant comorbidities were common complicating factors. The dominating modes of death were right heart failure and in patients with primary palliation a failing shunt circulation.