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Title: Hemodynamic assessment of pulmonary hypertension in corrected versus non-corrected grown-up congenital heart disease

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Body: PURPOSE: Pulmonary arterial hypertension (PAH) associated with congenital heart disease (CHD) is thought to result from persistent exposure of the pulmonary vasculature to increased blood flow of systemic-to-pulmonary shunts, and comprises a typical pulmonary arteriopathy in association with biventricular heart failure. Endothelial dysfunction has been demonstrated in individuals with congenital heart disease both before and after surgery. METHODS: 3107 right and left heart catheterizations were analyzed. Diagnoses were validated on the grounds of patient histories, imaging, clinical data and patho-anatomic evidence (2369 complete data sets). 241 data sets were from patients with CHD. RESULTS: Our database showed pre-tricuspid defects in 162 patients, post-tricuspid defects in 36 patients and complex lesions in 43 patients. 44 patients with CHD had undergone any correction. PH was observed in 27 patients with a correction status and in 22 patients with non-corrected CHD. Mean survival of “corrected” patients with PH (7.3 years) was worse than in “non-corrected” patients PH (11.4 years; p=0.009). There was a significant difference in age (p=0.05), mixed venous saturation (SvO2) (p=0.01), diastolic pulmonary artery pressure (dPAP) (p=0.018) and mean pulmonary capillary wedge pressure (mPCWP) (p=0.006) between “corrected” patients with PH and PH patients who did not undergo surgical or interventional correction. CONCLUSION: In an analysis accounting for confounders, grown-up patients with PH due to CHD after any corrective procedure show a worse survival than similar patients without correction.