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

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Body: PURPOSE: Pulmonary arterial hypertension (PAH) associated with congenital heart disease (CHD) is included in group 1 of the pulmonary hypertension (PH) clinical classification. The persistent exposure of the pulmonary vasculature to increased blood flow due to systemic-to-pulmonary shunts as well as increased pressure may result in a typical pulmonary arteriopathy that leads to an increase in invasively measured mean pulmonary arterial pressure (mPAP) ≥25mmHg at rest. 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). 257 data sets were from patients with CHD. RESULTS: Underlying diagnoses were: pre-tricuspid defects in 172 patients, post-tricuspid defects in 38 patients and complex lesions in 47 patients. Of the 257 patients with CHD (38 were corrected), 141 patients had normal hemodynamics (“Non-PH” mPAP<25mmHg). Of the remaining 116 patients with PH (with wedge tracings missing or insufficient in 19 cases), 51 qualified as pre-capillary PH (CHD-PAH; PCWP≤15mmHg), 46 had CHD with elevated left ventricular filling pressures (CHD-PH; mPCWP>15mmHg; Table 1).

Table 1. Hemodynamic data of patients with CHD and pulmonary hypertension

	CHD-PAH PCWP≤15mmHg (n=51)	CHD-PH PCWP>15mmHg (n=46)
age	51,3±17	60.6±15
sPAP (mmHg)	63.4±26.2	64.7±20.6
dPAP (mmHg)	26.1±11.5	26.9±9.2
mPAP (mmHg)	40,3±16,3	41.4±13.1
mPCWP (mmHg)	10.1±3.4	24.2±6.2

CONCLUSION: The data demonstrate that a significant proportion (almost 50%) of patients with PH in grown-up congenital heart disease suffer from post-capillary pulmonary hypertension.