PW1-10

Clinical classification of congenital heart disease associated pulmonary hypertension. Does it work for pediatrics? Analysis of the TOPP registry (Tracking outcome and practice in pediatric pulmonary hypertension)

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Objectives:
Current guidelines for pulmonary hypertension (PH) include a subclassification of congenital heart disease (CHD) into 4 groups: Eisenmenger (A), PAH-CHD associated to left to right shunt and high pulmonary vascular resistance(PVR) (B), PAH with small septal defects (C) and PAH after corrective surgery (D). We used a pediatric PH registry (TOPP) to evaluate the feasibility of classifying the pediatric patients.
Methods:
TOPP was designed before publication of these guidelines and did not use the PAH-CHD subclassification. However, specific details were collected to describe the anatomy, hemodynamics and surgical status. Two investigators reviewed independently all data and classified the patients. When agreement was obtained the patient was coded as A, B, C or D. If there was a discrepancy a consensus was reached by discussion. If consensus was not obtained,the patient was considered "not classified" (NC).(A) was defined as a large unrestrictive shunt defect with a Sa O_2<90%, (B) as a defect with SaO_2 >90% and indexed PVR >3 WU*m^2, (C) as small defects as described by the investigator or moderate defects and patient considered too young for the development of pulmonary vascular disease and (D) as PAH after complete repair .
Results:
Demographics, anatomy and indexed PVR are shown in the table. 27 patients diagnosed as idiopathic PAH had some type of CHD and were reclassified in (B) (3/27), (C) (19/27) and (D) (5/27).The largest group was PAH after corrective surgery 48/142 but a significant number of patients were not classified(NC) 18/142.

Group (n)	A (24)	B (25)	C (27)	D (48)	NC (18)
Age (years) Mean ((SD)	10.0 (6.3)	7.2 (5.9)	6.0 (5.4)	8.1 (4.9)	6.4 (4.9)
ASD	7	7	18	13	6
VSD	17	15	6	27	6
PDA	5	7	9	14	7
AVC	2	2	0	4	1
Other	3	5	3	3	4
PVRi WU*m2 Mean (SD)	21.0 (8.6)	13.2 (6.6)	14.4 (8.9)	17.1 (13.3)	15.5 (7.5)

Conclusions:
The subclassification of PAH-CHD seems feasible for pediatric patients in group A, B and D but was difficult to apply to group C with the current description of absolute defect size which does not always relate to children. The largest group was D, characterized by high PVR raising concerns about the appropriateness of surgical repair. Several participating sites classified patients with very small shunts as idiopathic PAH. In addition several patients with complex CHD or patients who never had shunts were difficult to classify. A more tailored description appears necessary in particular for small defects or complex forms of CHD.