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Title: Pulmonary hypertension associated with congenital heart disease (PH-CHD): Results from the ASPIRE registry

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Body: Introduction: Current guidelines describe 4 groups of PH-CHD: A Eisenmengers; B Large defect with left-right shunt; C Small defect; D Previously closed defect. There are sparse data comparing these groups. Methods: Data for all 198 PH-CHD pts in the ASPIRE (Assessing the Spectrum of Patients Identified at a REferral centre) registry were reviewed. Results: Although group B had milder haemodynamics and a lower proportion in functional class (FC) III or IV compared with groups C and D (Table) there was no significant difference in survival (Figure). In group A, Age, FC, DL_{CO} and incremental shuttle walking distance (ISWD) predicted survival at univariate, and ISWD at multivariate analysis, while survival in the 23% with trisomy-21 was not different to those without trisomy-21. Conclusions: Despite differing characteristics, survival of PH-CHD pts referred to specialist centres does not significantly differ between the 4 groups. Further details of medical and interventional therapies will be presented.

Group (n)	A (108)	B (49)	C (14)	D (27)
Age, yrs	37±15	47±17	44±17	48±19
WHO III/IV, %	62/11	47/6	46/31	54/8
ISWD, m	187±117	285±201	168±145	240±205
DLCO % pred	72±20	85±29	63±21	60±13
SaO2 %	83±8	94±4	91±6	94±3
PH Therapy %	81	55	100	81
sVO2 %		82±7	65±11	69±11
mRAP		11±5	9±5	12±6
mPAP		43±16	54±21	51±17

CI		3.7±0.6	2.6±0.8	3.0±0.7
PVR		313±165	955±270	667±499

(Data is presented as mean ± standard deviation, unless otherwise stated)