Cardiopulmonary Exercise Testing Predicts Outcome in Paediatric Pulmonary Hypertension

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INTRODUCTION
Pulmonary hypertension (PH) is associated with exercise intolerance and poor survival. Exercise limitation, measured by 6-minute walking distance tests (6MWDT) and cardiopulmonary exercise tests (CPET) has been robustly shown to correlate with disease severity and outcome in adult PH populations. Accordingly these measures have proved invaluable in guiding therapeutic decisions and as outcome measures in clinical trials. Data for their validity in children with PH are lacking. We aimed to analyse the relationship between CPET parameters and outcome in paediatric PH.

METHOD
A single center retrospective study of PH patients less than 18 years old that had undergone CPET between the dates April 2004 and April 2014. Exclusion from CPET was based upon height <120cm, or resting symptoms too severe for formal testing. Patients with cardiomyopathy or univentricular physiology were also excluded. All CPET and 6MWDT took place within the same 24-hour period. The need for ethical approval was waived.

RESULTS
93 patients underwent exercise testing. The median (IQR) age was 14.2 years (10.9-15.9), height 152.0cm (135.0-162.0) and weight 44.0kg (30.0-53.0). 46 (49.5%) had PH associated with congenital heart disease whilst 37 (39.8%) had idiopathic PH. WHO functional class was 1, 2, 3 and 4 in 16, 38, 38 and 1 patients respectively. Patients experienced significant exercise limitation; mean (SD) percentage predicted 6MWDT (6MWDT%) 57.7% (15.6%), peak VO2 (PVO2%) 54.9% (20.7%). Both 6MWD% and PVO2% correlated with WHO (r² 0.27 and 0.25 respectively, p<0.001) with a weaker correlation for VE/VCO2slope (r² 0.17, p<0.001). Over a median follow-up of 3.53 years (1.62-5.63), 16 patients died and 2 were transplanted. 6MWDT% did not predict outcome (HR 0.67, p=0.07) however, the following CPET measures were prognostic; PVO2%, VE/VCO2 slope; hazard ratio for each standard deviation increase of 0.52, 0.48 and 2.6 respectively.

CONCLUSION
In this large study we report for the first time the prognostic utility of cardiopulmonary exercise testing in children with pulmonary hypertension. Our findings support the use of CPET in paediatric PH.