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Survival adjusted with age is similar in adults with Eisenmenger syndrome and other types of pulmonary artery hypertension.

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Introduction : Patients with Eisenmenger syndrome (ES) are considered to carry a better prognosis than other pulmonary artery hypertension (PAH) causes because of a more favourable hemodynamic profile. However, patients with ES usually present younger than other patients with PAH. Hence, such comparison for survival is potentially biased if this is not adjusted for age. Finally, the survival of patients with PAH consecutive to late closure of congenital heart defect (CHD-PAH) is not clarified. We aimed to compare the survival adjusted for age in ES, CHD-PAH and other causes of PAH.
Methods: We retrospectively studied all patients with PAH followed in our tertiary centre since 2003. Events outcome were defined as death or transplantation.
Results: Out of 150 patients with PAH, 32 had ES (21%) and 11 had CHD-PAH (7%). Median follow-up was 51 (26 to 81) months. The median age at presentation was lower for patients with ES and CHD-PAH: 26 (7 to 46) and 38 (18 to 56) years, respectively, versus 54 (41 to 66) years for patients with other causes of PAH, ($p<0.001$ and 0.006 for ES and CHD-PAH, respectively). Events during follow-up occurred in 8 (25%) of ES, 2 (18%) of CHD-PAH and 34.7 % of patients with PAH. Patients with ES experienced significantly less events compared to patients with PAH (8 versus 48, $p=0.019$). Six patients with ES died, due to complication of cyanosis in 4 cases. The outcome for patients with CHD-PAH was similar than patients with other causes of PAH ($p=0.3$). Crude hazard ratio of events in ES and CHD-PAH population compared to patients with other causes of PAH was 0.3 (0.1 to 0.8) and 0.4 (0.1 to 1.8), respectively. When adjusted for age, ES and other causes of PAH experienced a similar longevity (0.6 range 0.2 to 1.9).
Conclusion: The longevity is similar between ES, PAH-CHD and other types of PAH. The long-term complications of cyanotic heart disease may counterbalance the relative protection of ES for right ventricular dysfunction.