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CLINICAL RESEARCH

Outcome of adults with Eisenmenger syndrome treated with drugs specific to pulmonary arterial hypertension: A French multicentre study

Devenir des adultes avec syndrome d'Eisenmenger traités par médicaments spécifiques anti-hypertenseur pulmonaire : données d'une étude multicentrique française

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Abbreviations: CHD, Congenital heart disease; ERA, Endothelin receptor antagonist; ES, Eisenmenger syndrome; MCE, Major clinical event; NYHA/WHO FC, New York Heart Association/World Health Organization functional class; PAH, Pulmonary arterial hypertension; PAH-SDT, Pulmonary arterial hypertension-specific drug therapy; SaO_2, Peripheral arterial oxygen saturation.

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KEYWORDS
Eisenmenger syndrome;
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Summary
Background. — The relationship between pulmonary arterial hypertension-specific drug therapy (PAH-SDT) and mortality in Eisenmenger syndrome (ES) is controversial.
Aims. — To investigate outcomes in patients with ES, and their relationship with PAH-SDT.
Methods. — Retrospective, observational, nationwide, multicentre cohort study.
Results. — We included 340 patients with ES: genetic syndrome ($n = 119$; 35.3%); pretricuspid defect ($n = 75$; 22.1%). Overall, 276 (81.2%) patients received PAH-SDT: monotherapy (endothelin receptor antagonist [ERA] or phosphodiesterase 5 inhibitor [PDE5I]) 46.7%; dual therapy (ERA + PDE5I) 40.9%; triple therapy (ERA + PDE5I + prostanoid) 9.1%. Median PAH-SDT duration was 5.5 years [3.0—9.1 years]. Events (death, lung or heart-lung transplantation) occurred in 95 (27.9%) patients at a median age of 40.5 years [29.4—47.6]. The cumulative occurrence of events was 16.7% [95% confidence interval 12.8—21.6%] and 46.4% [95% confidence interval 38.2—55.4%] at age 40 and 60 years, respectively. With age at evaluation or time since PAH diagnosis as time scales, cumulative occurrence of events was lower in patients taking one or two PAH-SDTs ($P = 0.0001$ and $P = 0.004$, respectively), with the largest differences in the post-tricuspid defect subgroup ($P < 0.001$ and $P < 0.02$, respectively) versus patients without PAH-SDT. By multivariable Cox analysis, with time since PAH diagnosis as time scale, New York Heart Association/World Health Organization functional class III/IV, lower peripheral arterial oxygen saturation and pretricuspid defect were associated with a higher risk of events ($P = 0.002$, $P = 0.01$ and $P = 0.04$, respectively), and one or two PAH-SDTs with a lower risk of events ($P = 0.009$).

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