

Impact of Cardiopulmonary Comorbidities on Survival in Idiopathic Pulmonary Arterial Hypertension Patients: Data from the Hellenic pulmonary hypertension registry (HOPE)

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Background

IPAH patients with cardiopulmonary comorbidities, have higher mortality risk and correspond less well to PAH medication. However, the influence of each of these on survival has not been fully established.

Methods

The Hellenic Pulmonary Hypertension Registry (HOPE) registry contains patients from all PH groups in Greece. This study focuses on IPAH patients with cardiopulmonary comorbidities.

Results

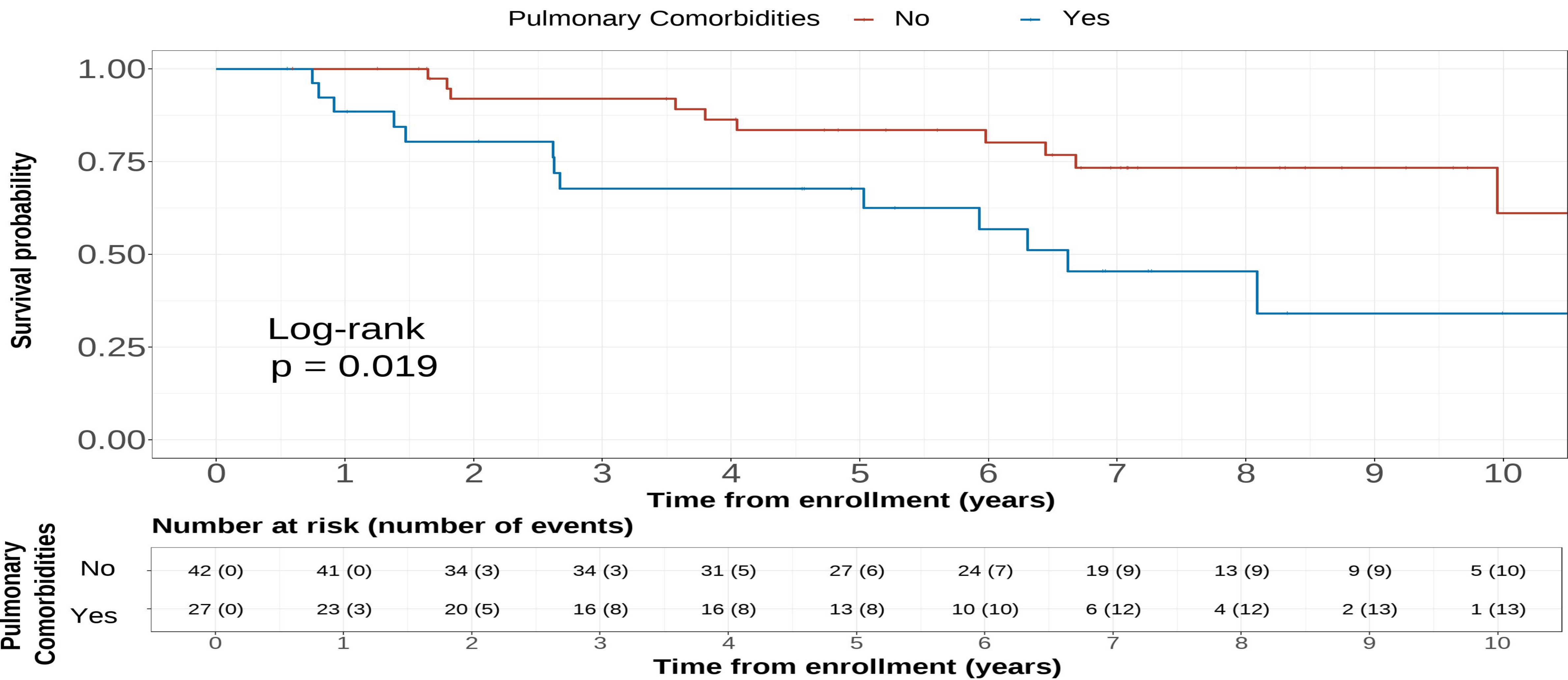
- 69 IPAH patients with comorbidities were included (71% female), median (IQR) age at first visit 60 (15) years.
- After a median follow up of 5.9 years, 25 deaths (36.2%) occurred (6 deaths per hundred patient-years).
- The presence of at least one pulmonary comorbidity caused a significant increase in mortality (Table 1).
- 5-year survival rate of IPAH patients with pulmonary comorbidities was 67.7% compared to 83.5% of the patients without pulmonary comorbidities (p=0.019, Figure).

Table 1. Univariate and multivariate Cox regression analysis

Characteristic	Univariate				Multivariate			
	N	HR [†]	95% CI [†]	p-value	HR [†]	95% CI [†]	p-value	
Obesity	69	1.81	0.67, 4.85	0.2	1.88	0.70, 5.08	0.2	
Coronary artery disease	45	0.94	0.12, 7.26	>0.9				
Hypertension	57	0.99	0.40, 2.44	>0.9				
Diabetes	45	0.41	0.05, 3.15	0.4				
Pulmonary Comorbidities	69	2.54	1.13, 5.71	0.024	2.60	1.16, 5.82	0.021	

[†] HR = Hazard Ratio, CI = Confidence Interval

Survival Analysis of PAH patients with cardiovascular comorbidities depending on the presence of pulmonary comorbidities



Conclusions

The presence of pulmonary comorbidities in IPAH is a major risk factor for mortality. Other conditions associated with an increased risk of left ventricular diastolic dysfunction do not seem to affect survival.