

**Table 1:** Clinical and hemodynamic data, according to the time of diagnosis of pulmonary hypertension

	<b>Before July 2007 N= 861</b>	<b>After July 2007 N= 167</b>
<b>Age (yr)</b>	45 ± 17	51 ± 18*
<b>Gender (%male)</b>	29%	26%
<b>Time symptoms- diagnosis (yr)</b>	3.7 ± 6.3	2.2 ± 4.7*
<b>WHO FC III-IV</b>	69%	68%
<b>mPAP (mmHg)</b>	56 ± 16	48 ± 13*
<b>CI (l/min/m<sup>2</sup>)</b>	2.6 ± 0.8	2.8 ± 1.0
<b>PVR (WU)</b>	12 ± 7	10 ± 6*
<b>RAP (mmHg)</b>	8.6 ± 5.2	9.6 ± 6.1
<b>6MWD (m)</b>	360 ± 117	378 ± 135

\*p<0.05 compared with the group diagnosed before July 2007

**Definition of abbreviations:** WHO-FC: functional classification according to World Health Organization mPAP: mean pulmonary artery pressure. PVR: pulmonary vascular resistance. CI: cardiac index. RAP: right atrial pressure. 6MWD: 6-minute walk distance.

**All patients**

**Patients with IPAH**

	<i>HR [CI 95%]</i>	<i>p-value</i>	<i>HR [CI 95%]</i>	<i>p-value</i>
<b>Age (10 years)</b>	1.15 (1.08-1.23)	<0.001	1.08 (0.95-1.23)	0.234
<b>Gender (men)</b>	1.27 (1.00-1.60)	0.045	2.96 (1.96-4.53)	<0.001
<b>6 MWT (20m)</b>	0.93 (0.91-0.95)	<0.001	0.92 (0.88-0.96)	<0.001
<b>WHO-FC</b>	1.65 (1.38-1.98)	<0.001	2.90 (2.02-4.18)	<0.001
<b>Pericardial effusion</b>	1.34 (1.01-2.10)	0.042	1.1 (0.85-1.6)	0.364
<b>Year of diagnosis</b>		0.717		0.506
<b>1998- 2001</b>	reference	-	reference	-
<b>2002-2004</b>	1.12 (0.85-1.48)	0.428	0.74 (0.44-1.26)	0.269
<b>2005-2008</b>	1.04 (0.76-1.42)	0.831	0.80 (0.44-1.43)	0.443
<b>CO ( l/min)</b>	0.76 (0.69-0.83)	<0.001	0.71 (0.58-0.88)	0.001
<b>mPAP (5 mm Hg)</b>	1.00 (0.97-1.04)	0.912	1.12 (1.04-1.20)	0.002
<b>PVR (5 UW)</b>	1.15 (1.07-1.24)	<0.001	1.25 (1.07-1.45)	0.004
<b>RAP (5 mm Hg)</b>	1.40 (1.27-1.56)	<0.001	1.81 (1.43-2.28)	<0.001
<b>IC ( l/min/m<sup>2</sup>)</b>	0.60 (0.50-0.72)	<0.001	0.49 (0.33-0.72)	<0.001
<b>DLCO (10)</b>	0.83 (0.78-0.89)	<0.001	0.84 (0.75-0.94)	0.002
<b>BP (10 mm Hg)</b>	0.98 (0.93-1.04)	0.552	0.93 (0.83-1.04)	0.203
<b>HR (10 )</b>	1.08 (0.97-1.20)	0.179	1.36 (1.12-1.65)	0.002
<b>Subtypes of PH</b>		<0.001		
<b>Idiopathic</b>	reference.	-		
<b>CTD</b>	1.74 (1.25-2.42)	0.001		
<b>CHD</b>	0.69 (0.47-1.01)	0.057		
<b>CTEPH</b>	0.98 (0.66-1.45)	0.911		
<b>HIV</b>	0.91 (0.51-1.62)	0.738		
<b>PoPH</b>	2.00 (1.25-3.13)	0.004		
<b>TOS</b>	0.87 (0.49-1.53)	0.616		
<b>PVOD</b>	4.18 (1.69-10.3)	0.002		
<b>2 or more etiologies</b>	1.00 (0.64-1.57)	0.991		
<b>others</b>	1.82 (1.06-3.11)	0.029		

**Table 2:** Univariate predictors of death in Cox's proportional hazards analysis  
**Definition of abbreviations:** IPAH: idiopathic PAH WHO-FC: functional classification according to World Health Organization. 6MWD: 6-minute walk distance. WHO-FC: functional classification according to World health organization. mPAP: mean pulmonary artery pressure. PVR: pulmonary vascular resistance. WU: Wood Units. CI: cardiac index. RAP: right atrial

pressure. CI: cardiac index. HR: Heart rate. PH: pulmonary hypertension. CTD: connective tissue disease. CHD: congenital heart disease. CTEPH: chronic thromboembolic pulmonary hypertension. HIV: human immunodeficiency virus. PoPH: portopulmonary hypertension. TOS: Toxic oil syndrome. PVOD: pulmonary veno-occlusive disease.

**Table 3:** Observed survival in current PAH registries from time of diagnosis

	REHAP	PHC	French Registry	REVEAL
<b>Patients with PAH</b>				
1 year	86%	86%	87%	88%
3 years	75%	69%	67%	72%
<b>Patients with IPAH</b>				
1 year	89%	92%	83%	
3 years	77%	75%	58%	

**Definition of abbreviations:** PAH: pulmonary arterial hypertension. IPAH: idiopathic pulmonary arterial hypertension. REHAP: Spanish Registry of Pulmonary Arterial Hypertension PHC: Pulmonary Hypertension Connection. REVEAL: Registry to Evaluate Early and Long-Term Pulmonary Arterial Hypertension Disease Management.