

**Supplementary Table S2** Events related to pulmonary arterial hypertension and death

	PAH-CTD		PAH-SSc		PAH-SLE		PAH-MCTD/CTD-other	
	Placebo N=167	Selexipag N=167	Placebo N=93	Selexipag N=77	Placebo N=37	Selexipag N=45	Placebo N=37	Selexipag N=45
<b>Primary composite endpoint of morbidity/mortality up to the end of treatment</b>	<i>Number of patients (%)</i>		<i>Number of patients (%)</i>		<i>Number of patients (%)</i>		<i>Number of patients (%)</i>	
All events	73 (43.7)	48 (28.7)	46 (49.5)	25 (32.5)	13 (35.1)	11 (24.4)	14 (37.8)	12 (26.7)
Hospitalisation for worsening of PAH	36 (21.6)	21 (12.6)	22 (23.7)	10 (13.0)	4 (10.8)	4 (8.9)	10 (27.0)	7 (15.6)
Disease progression	29 (17.4)	11 (6.6)	18 (19.4)	8 (10.4)	8 (21.6)	3 (6.7)	3 (8.1)	0
Death from any cause	3 (1.8)	12 (7.2)	1 (1.1)	4 (5.2)	1 (2.7)	3 (6.7)	1 (2.7)	5 (11.1)
Initiation of parenteral prostanoid therapy or long-term O <sub>2</sub> therapy for worsening PAH	5 (3.0)	3 (1.8)	5 (5.4)	2 (2.6)	0	1 (2.2)	0	0
Need for lung transplantation or BAS for worsening of PAH	0	1 (0.6)	0	1 (1.3)	0	0	0	0
<b>Secondary endpoint of all-cause death up to the end of the study</b>	<i>Number of patients (%)</i>		<i>Number of patients (%)</i>		<i>Number of patients (%)</i>		<i>Number of patients (%)</i>	
Death from any cause	34 (20.4)	33 (19.8)	22 (23.7)	17 (22.1)	7 (18.9)	4 (8.9)	5 (13.5)	12 (26.7)

BAS: balloon atrial septostomy; CTD: connective tissue disease; MCTD: mixed connective tissue disease; PAH: pulmonary arterial hypertension; SLE: systemic lupus erythematosus; SSc: systemic sclerosis.