Supplementary Table 1. Baseline, Week 17 and change from baseline to Week 17 in pulmonary vascular resistance (per protocol and all-treated set)

	Mean values ± SD at baseline		Mean values ± SD at Week 17		Mean change ± SD from baseline to		Treatment effect*
					We	ek 17	[95% CL;
	Placebo	Selexipag	Placebo	Selexipag	Placebo	Selexipag	Wilcoxon P-
							value]
Per protocol set							
N	6	29	6	29	6	29	
Pulmonary vascular	826.8±195.8	951.9±434.5	964.0±247.9	783.8±393.2	137.2 ±	−168.1 ±	-30.3%
resistance, dyn $\cdot$ s $\cdot$					84.9	241.6	[-44.7,
cm <sup>-5</sup>							-12.2;
							0.0045]
All-treated set							
N	10	32	10	32	10	32	
Pulmonary vascular	867.2 ±	948.6 ±	1090.8 ±	818.8 ±	223.6 ±	-129.8 ±	-33.0%
resistance, dyn · s ·	379.3	428.0	421.3	416.9	355.4	309.7	[-47.0,
cm <sup>-5</sup>							-15.2;
							0.0022]

<sup>\*</sup>Treatment effect calculated at Week 17 as the change in the geometric mean expressed as a percentage of the baseline value SD; standard deviation

## Supplementary Table 2. Serious adverse events during the study (safety set)

Serious adverse events	Placebo	Selexipag
	n=10	n=33
Patients with ≥1 serious		_
adverse event, n	4	6
Total number of serious	10	13
adverse events		
Headache	-	2
Pulmonary arterial	2	1
hypertension		
Dyspnoea	1	1
Chest pain	-	1
Myalgia	-	1
Nasopharyngitis	-	1
Nausea	-	1
Oral papilloma	-	1
Psychotic disorder	-	1
Sciatica	-	1
Urinary tract infection	-	1
Vomiting	-	1
Alcohol interaction	1	-
Benign prostatic hyperplasia	1	-
Cardiac failure	1	-

Cataract operation	1	-	
Chronic obstructive	1)	-	
pulmonary disease			
Respiratory infection	1	-	
Urinary retention	1	-	

## Supplementary Table 3. Prevalence of selected treatment-emergent adverse events over time in selexipag-treated patients

Adverse events	≥ <b>0</b> –27 days	<b>≥28</b> –55	<b>≥56</b> –83	≥84 days
(AE)*	(n=33)	days	days	(n=31)
		(n=32)	(n=32)	
Patients with ≥1 AE, n	26 (78.8)	24 (75.0)	22 (68.8)	21 (67.7)
(%)				
Different AEs, n	56	47	43	34
AE, n (%)				
Headache	21 (63.6)	16 (50.0)	16 (50.0)	14 (45.2)
Pain in extremity	10 (30.3)	9 (28.1)	9 (28.1)	4 (12.9)
Pain in jaw	8 (24.2)	8 (25.0)	6 (18.8)	6 (19.4)
Nausea	8 (24.2)	6 (18.8)	5 (15.6)	4 (12.9)
Flushing	4 (12.1)	4 (12.5)	4 (12.5)	4 (12.9)
Diarrhoea	5 (15.2)	4 (12.5)	3 (9.4)	2 (6.5)

<sup>\*</sup>Adverse events commonly associated with prostacyclin analogues