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Risk Assessment at Baseline and One Year In Patients With Pulmonary Arterial Hypertension (PAH): Data From the First 250 Patients Enrolled in SPHERE (Uptravi® [SelexiPag]: tHe UsErs dRug rEgistry)

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Background: Selexipag, a selective oral IP prostacyclin receptor agonist, is approved to delay disease progression and reduce the risk of hospitalization in patients with PAH. Use of routine risk assessment is recommended to guide treatment decisions in PAH and to monitor disease course over time. We assessed risk in the first 250 patients enrolled in SPHERE at baseline and at 1-year follow-up.

Methods: Initiated in November 2016, SPHERE is an ongoing US-based observational registry collecting real-world data from PAH patients treated with selexipag. Patients newly initiated on selexipag, or previously initiated and with a documented titration regimen are eligible. We assigned risk in a similar manner as the COMPERA and Swedish registries (Hoeper Eur Resp J 2017; Kylhammar Eur Heart J, 2017) and included at least one of the following variables: WHO or NYHA functional class, 6-minute walk distance, NT-proBNP, BNP, right atrial pressure, cardiac index, and mixed venous oxygen saturation. If both NT-proBNP and BNP values were available, both were used. A risk score (1=low risk, 2=intermediate risk, and 3=high risk) was assigned to each variable according to ESC/ERS guidelines thresholds. These values were averaged and rounded to the nearest integer to calculate a risk category for each patient of 1 (low risk), 2 (intermediate risk), or 3 (high risk). The time of selexipag initiation was considered baseline for this analysis.

Results: The data cut-off was August 21, 2018. At selexipag initiation, 58 (23.2%), 169 (67.6%), and 19 (7.6%) patients were classified as low-, intermediate-, and high-risk, respectively (Table). Median age was higher in intermediate- and high-risk groups. During follow-up, twenty-nine patients (11.6%) died: 2 (3.4%) from the low-risk group, 19 (11.2%) from the intermediate-risk group, and 7 (36.8%) from the high-risk group (risk could not be assigned for one patient who died). Selexipag maintenance doses and time to maintenance doses were similar in low-, intermediate- and high-risk groups (1400, 1200, and 1400 µg BID; 8.7, 8.3, and 8.3 weeks, respectively). Among patients who had risk assessment at both baseline and 1 year (196 patients or 78% of the total population), 22% had improved, 15% had worsened, and 63% had no change in risk group from baseline (see table).

Conclusions: Of patients with available data at baseline and 1 year, 85% remained in the same risk group (63%) or improved to a less severe risk group (22%).

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Figure 1. Disease Characteristics at Baseline (Selexipag Initiation) and Patient Disposition

Disease Characteristics at Baseline (Selexipag Initiation)[a] and Patient Disposition				
	All Patients (N=250)	**Low Risk (n=58)**	**Intermediate Risk (n=169)**	**High Risk (n=19)**
Disease characteristics				
Age (years)				
Median	60	48	62	64
Q1, Q3	49, 69	37, 61	54, 70	56, 72
Time from PAH diagnosis to selexipag initiation (years)				
Median	3.7	4.2	3.5	2.4
Q1, Q3	1.5, 7.5	1.5, 9.3	1.6, 7.5	0.4, 4.0
Etiology, n (%)				
PAH associated with other disease state	94 (37.6)	16 (27.6)	72 (42.6)	5 (26.3)
Idiopathic PAH	131 (52.4)	35 (60.3)	82 (48.5)	12 (63.2)
Other or unknown	25 (10.0)	7 (12.1)	15 (8.9)	2 (10.5)
NYHA/WHO FC, n (%)				
I	11 (4.4)	4 (6.9)	7 (4.1)	0 (0)
II	69 (27.6)	39 (67.2)	30 (17.8)	0 (0)
III	138 (55.2)	9 (15.5)	119 (70.4)	10 (52.6)
IV	8 (3.2)	0 (0.0)	3 (1.8)	5 (26.3)
Unknown	24 (9.6)	6 (10.3)	10 (5.9)	4 (21.1)
Patient disposition				
Duration of selexipag treatment from initiation to study enrollment (months)				
Median	7.1	8.0	7.1	6.4
Q1, Q3	2.0, 11.3	3.6, 12.1	1.9, 11.0	1.3, 11.6
Total duration of selexipag treatment since initiation (months)				
Median	18.2	20.0	17.9	13.6
Q1, Q3	12.1, 24.5	15.2, 25.7	11,7, 24.2	7.2, 25.0
Selexipag maintenance dose[b] (µg BID)				
Median	1200	1400	1200	1400
Q1, Q3	800, 1600	800,1600	800, 1600	1000, 1600