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Characteristics and outcomes of patients with pulmonary arterial hypertension transitioning to selexipag from another prostacyclin pathway agent in the SPHERE registry

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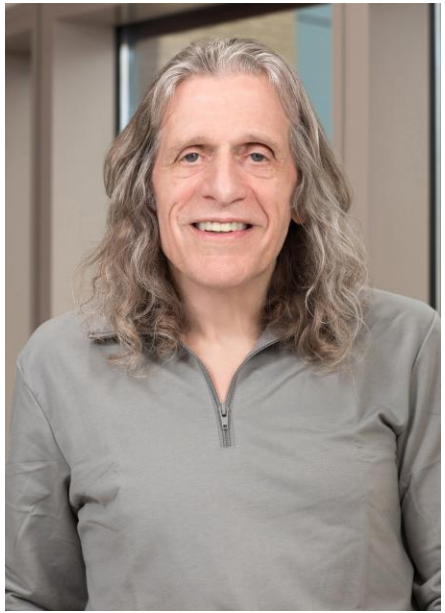

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Disclosures: I am a speaker for Bayer (non-branded) and a scientific advisory board member for Actelion, a Johnson & Johnson Company, Acceleron (Merck), Aerovate, Aerami, Roivant, and United Therapeutics

Objective

- To describe the characteristics and outcomes of patients with pulmonary arterial hypertension who transitioned to selexipag from another prostacyclin pathway agent, using data from the real-world US SelexiPag: tHe usErs dRug rEgistry (SPHERE)

SelexiPag: tHe usErs dRug rEgistry (SPHERE)

N=759
Adults with PAH

- US, multicenter, prospective, real-world, observational selexipag drug registry
- Followed for up to 18 months
- NCT03278002

Data collected at routine clinical visits and analysis:

- Patient demographics
- Medical history
- Disease characteristics
- WHO functional class
- REVEAL 2.0
- Prior PAH therapy (past 12 months)
- Selexipag dose regimens and titration
- Selexipag discontinuation and reason
- Time to first hospitalization
- Overall survival
- Safety

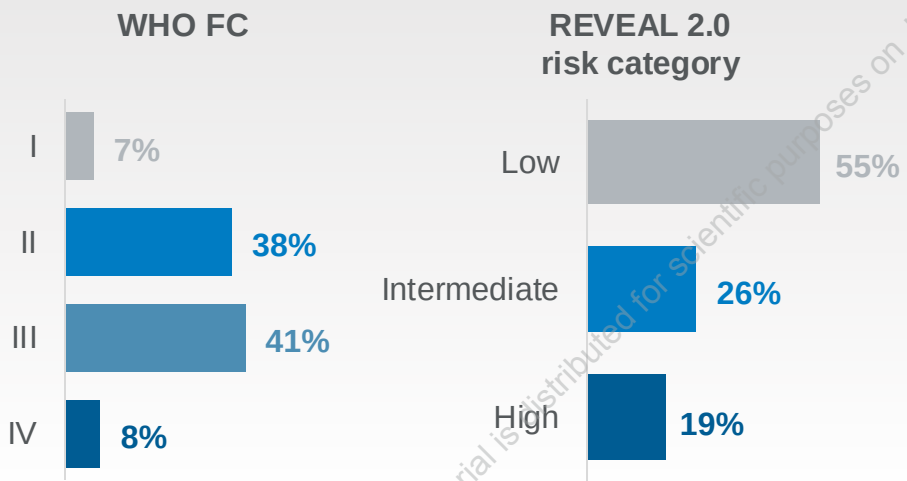
WHO, world health organization; PAH, pulmonary arterial hypertension

SPHERE registry: Patients with PAH transitioning from a different PPA to selexipag

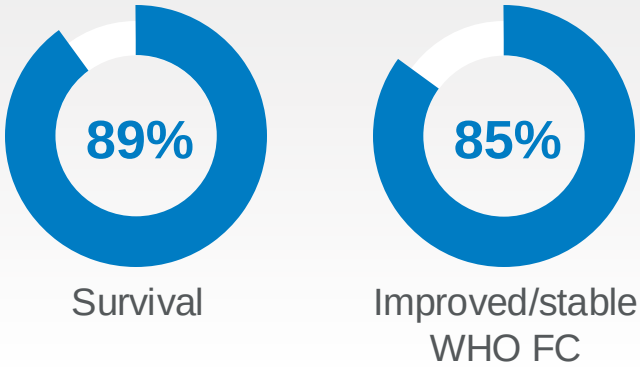
PPA → Selexipag

124 (16%) patients in the SPHERE registry were treated with a different PPA prior to starting selexipag

Patients who transitioned were:



Carefully selected patients may be able to successfully transition to selexipag and reach stable or improved disease at 18 months:



FC, functional class; PAH, pulmonary arterial hypertension; PPA, prostacyclin pathway agent;
REVEAL, Registry to Evaluate Early and Long-Term PAH Disease Management; SPHERE, SelexiPag: tHe usErs dRug rEgistry; WHO, World Health Organization.

124 patients with PAH in the SPHERE registry transitioned from another PPA to selexipag

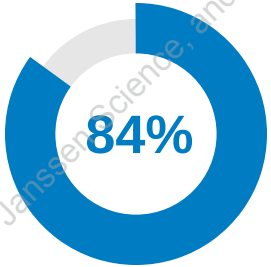
Total patients with PAH in SPHERE (N=759)

Transitioned from another PPA to selexipag (n=124 [16%])

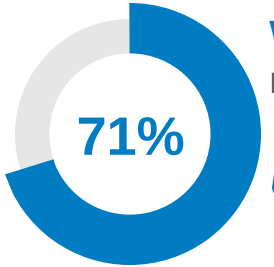
From inhaled PPA (n=58)	From oral PPA (n=14)	From IV/SC PPA (n=52)
47%	11%	42%

Patients who completed 18 months of observation (n=108; 87.1%)

Patients who transitioned to selexipag

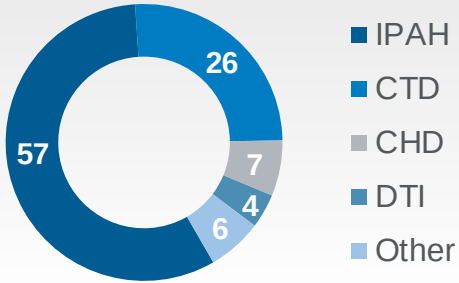


Female
n=104



White race
n=88

PAH etiology (%)



Median age
59 years
(IQR 48.5-65.0)

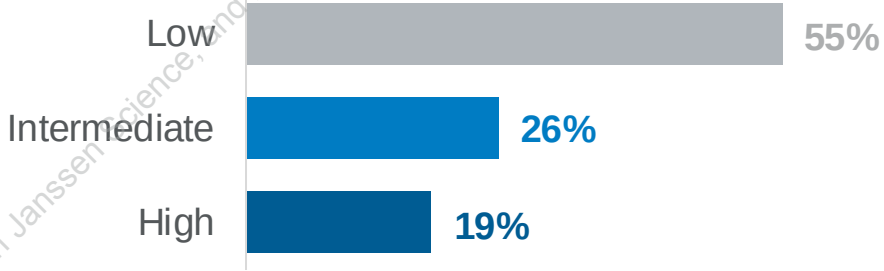
Median time from diagnosis to selexipag initiation
5.8 years
(IQR 0.1-35.5)

CHD, congenital heart disease; CTD, connective tissue disease; DTI, drug and toxin induced; IPAH, idiopathic pulmonary arterial hypertension; IQR, interquartile range; IV, intravenous; PAH, pulmonary arterial hypertension; PPA, prostacyclin pathway agent; SC, subcutaneous; SPHERE, SelexiPag: tHe usErs dRug rEgistry.

Baseline characteristics of patients who transitioned to selexipag

WHO functional class

REVEAL 2.0 risk category



Median 6MWD
324.0 m
(IQR 223.5-420.0)

Median PVR
5.1 Wood units
(IQR 3.6-9.0)

Median mPAP
42 mmHg
(IQR 32-55)

Median BNP/ NT-proBNP
52 ng/L (IQR 33-160)/
284 ng/L (IQR 96-1071)

6MWD, 6-minute walk distance; BNP, brain natriuretic peptide; IQR, interquartile range; mPAP, mean pulmonary artery pressure; NT-proBNP, N-terminal prohormone of brain natriuretic peptidePVR, pulmonary vascular resistance; REVEAL, Registry to Evaluate Early and Long-Term PAH Disease Management; RHC, right heart catheterization; WHO, World Health Organization.

Patient outcomes after transitioning to selexipag

Parameter[a]	All transitioned patients (n=124)	From inhaled (n=58)	From oral (n=14)	From IV/SC (n=52)
Overall survival at 18 months, % of patients (95% CI)	89 (82-94)	90 (78-95)	93 (59-99)	SC: 100 (100-100) IV: 86 (72-94)
WHO functional class change at 18 months, n/N[b] (%)				
Improved	12/58 (21)	7/28 (25)	1/7 (14)	4/23 (17)
Stable	37/58 (64)	17/28 (61)	5/7 (71)	15/23 (65)
Worsened	9/58 (16)	4/28 (14)	1/7 (14)	4/23 (17)
Reason for permanent selexipag discontinuation, n (%)				
Death	8 (6)	3 (5)	0	2 (4)
AE	24 (19)	11 (19)	0	10 (19)
AE related to PAH progression	11 (9)	5 (9)	0	4 (8)
Selexipag-related AE	12 (10)	5 (9)	0	5 (10)
Any other reason	11 (9)	5 (9)	1 (7)	5 (10)

Median selexipag maintenance dose among all transitioned patients was 1400 µg twice daily

[a]Numbers may not add to 100% due to rounding; [b]Patients with evaluable data at 18 months.
AE, adverse event; CI, confidence interval; IV, intravenous; PAH, pulmonary arterial hypertension; SC, subcutaneous; WHO, World Health Organization.

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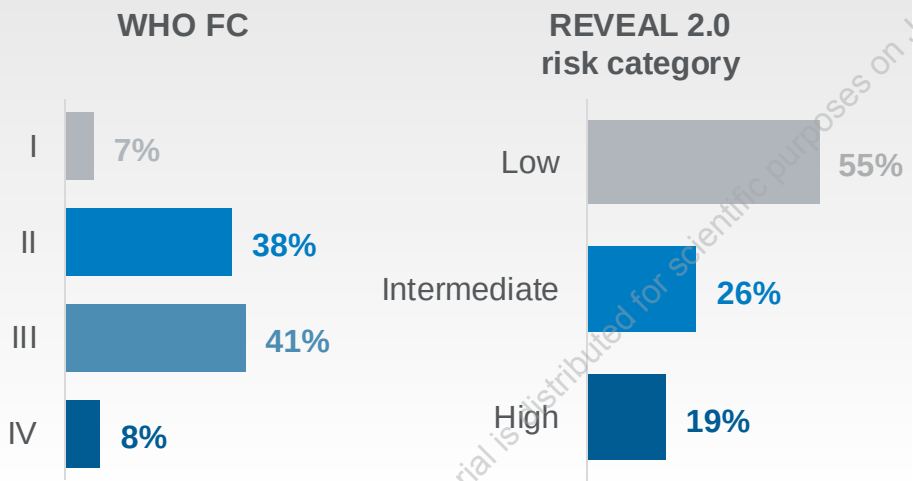

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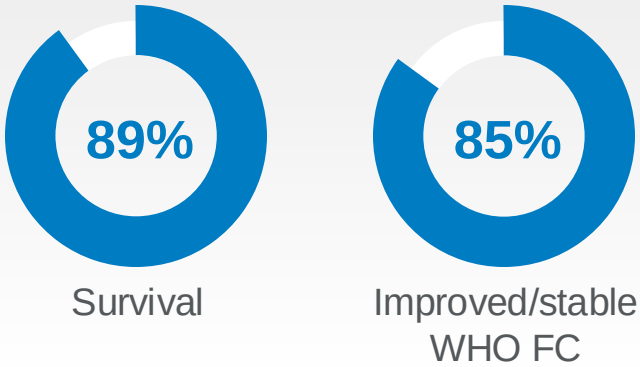

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