

Impact Of Functional Class Change On Survival In Patients With Pulmonary Arterial Hypertension In The Reveal Registry

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Purpose: The Registry to Evaluate Early And Long-term Pulmonary Arterial Hypertension (PAH) Disease Management (REVEAL), a 55-center observational, US-based study, provides information on current demographic, clinical, and treatment patterns in patients with PAH. To determine if patients who improve from functional class (FC) III at enrollment to FC I/II at follow-up assessment have a better survival rate compared with patients who remain stable FC III, two-year outcomes were analyzed.

Methods: 1,082 adult patients who were enrolled in REVEAL and were assessed as FC III at enrollment (based on the most recent pre-enrollment evaluation) and had at least one follow-up FC assessment within the first year after enrollment were included in these analyses. We classified patients as: 1) improved if FC III improved to FC I/II; 2) stable if remained FC III; or 3) deteriorated if worsened to FC IV (based on their first FC follow-up assessment after enrollment). We compared the survival rates (estimates +/- SE) of these three subgroups from the time of the first follow-up FC assessment based on the log-rank test.

Results: Demographics and clinical characteristics of FC III patients at enrollment who improved to FC I/II, remained stable at FC III, or deteriorated at their first follow-up assessment are presented in the Table. Twenty-six percent (n = 281) improved, 66% (n = 718) remained stable and 8% (n = 83) deteriorated. Kaplan-Meier survival from first follow-up FC assessment in these subgroups are presented in the Figure; two-year survival rates were 88 ± 2%, 76 ± 2%, and 34 ± 6%, for FC I/II, FC III, and FC IV patients, respectively (P <0.001 for all pairwise comparisons). Results are similar for both newly and previously diagnosed patients (i.e., confirmatory right heart catheterization within or not within 3 months prior to enrollment) who were FC III at enrollment.

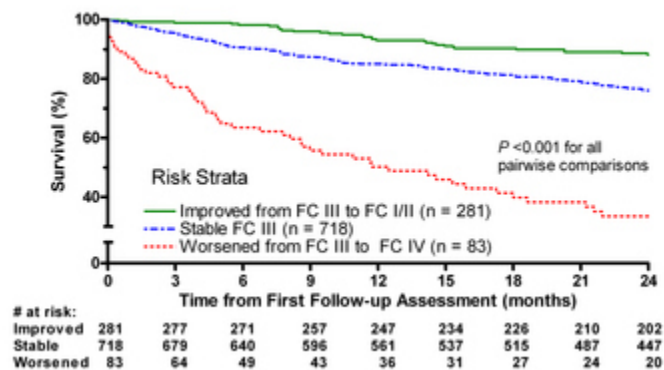
Conclusions: These results suggest that patients who improve from FC III to FC I/II have a better two-year survival compared with patients who remain FC III.

Table. Demographics and Characteristics of FC III Patients

Characteristic	Improved FC III FC I/II N=281	Stable FC III N=718	Worsened FC III IV N=83
Mean (SD) age at enrollment, ^a years	50 (16)	54 (16)	55 (16)
Newly diagnosed at enrollment, ^b n (%)	98 (35)	193 (27)	23 (28)
Female gender, ^c n (%)	224 (80)	577 (80)	70 (84)
Group I PAH Diagnosis at Enrollment,^c n (%)			
Idiopathic	131 (47)	353 (49)	41 (49)
Familial	11 (4)	19 (3)	3 (4)
Pulmonary Veno-Occlusive Disease	1 (0.4)	3 (0.4)	1 (1.2)
Associated with:			
Connective Tissue Disease	69 (25)	204 (28)	23 (28)
Congenital Heart Disease	32 (11)	65 (9)	8 (10)
Portal Hypertension	14 (5)	33 (5)	1 (1)
Drugs and Toxins	18 (6)	27 (4)	6 (7)
HIV	4 (1)	5 (1)	0
Other	1 (0.4)	9 (1)	0
Mean (SD) 6-minute walk distance at enrollment, ^a (m)	365 (102)	310 (112)	265 (114)
Mean (SD) 6-minute walk distance at 1 st follow-up FC assessment, ^a (m)	393 (101)	318 (103)	204 (96)
Mean (SD) change in 6-minute walk distance (m) between enrollment and 1 st follow-up FC assessment ^a	28 (73)	-1 (68)	-32 (83)

Mean (SD) right atrial pressure (mRAP) at enrollment, ^c mm Hg	9 (5)	9 (6)	10 (5)
Mean (SD) cardiac index at enrollment, ^c L/min x m ²	2.4 (1.0)	2.4 (0.8)	2.3 (0.8)
^a P <0.001 improved vs. stable ^b P = 0.012 improved vs. stable ^c P = NS improved vs. stable			

Figure. Two-year survival from first follow-up FC assessment of patients who were FC III at enrollment



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