

Frequency Of Palliative Care Referrals In Newly Diagnosed Pulmonary Arterial Hypertension (PAH): The Pulmonary Hypertension Association Registry

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Background: In addition to a high mortality, PAH imposes significant morbidity. Despite improvements in functional capacity (FC) and delayed time-to-clinical-worsening in randomized clinical trials, changes in symptoms and health-related quality of life (HRQOL) are of limited magnitude and rarely achieve clinical significance. Palliative care (PC) can improve HRQOL, decreases anxiety and depression, and may improve survival in some patients with chronic and terminal disease. However, there are no studies to date examining the use of PC in a PAH population. Therefore, we sought to determine the frequency of PC referrals in the PHA Registry (PHAR), and to characterize PAH patients referred to PC.

Methods: We performed a cohort study of newly-evaluated patients ≥18 years of age with PAH. Baseline demographic, socioeconomic status (SES), clinical phenotype, and supportive care data were collected, as well as the SF-12 and emphasis-10. Time-to-PC-referral was determined by calculating the time from enrollment to reported PC referral.



Results: We included 487 newly diagnosed PAH patients in the analysis. Initial and subsequent assessment of PC referral status were made at follow-up visit. 25 (5.1%) were referred to PC during follow-up (average follow-up for PHAR patients was 13.5 months (IQR 7.7, 19.8). The median time to PC referral for those referred was 9.3 months (IQR 5.2, 17.4) after initial evaluation. PC referred patients were older, with lower BMI, shorter 6-minute walk distance, and poor hemodynamics. They were more frequently diagnosed with CTD-APAH, more frequently on parenteral prostacyclin and supplemental oxygen therapy, and reported a worse HRQOL by EmPHasis-10 and SF-12 Mental Health Composite Scale score at the time of enrollment (Table 1).

Conclusions: Incident adult PAH patients enrolled in PHAR had a very low rate of referral to PC during early-to-immediate-term follow-up. PC referred patients were more likely to be receiving parenteral therapy and be on supplemental oxygen, and to report a worse HRQOL by EmPHasis-10 and SF-12 Mental Health Composite score at the time of enrollment. These findings demonstrate the need for further study into the role and timing of PC in PAH and to identify barriers to PC referral.



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Figure 1. Table 1: Characteristics of Patients Referred to Palliative Care in the PHA Registry

TABLE 1: Characteristics of Patients Referred to PC

Type	Baseline Characteristics	No PC Referral (n=413) No. (%) or Median (IQR)	PC Referral (n=25) No. (%) or Median (IQR)
Demographics	Age, years	56.6 (42.6, 67.3)	71.1 (47.6, 74.4)
	Female	304 (73.6)	21 (84.0)
	Race		
	White	290 (73.8)	19 (79.2)
	Black	49 (12.5)	2 (8.3)
	Other	30 (7.6)	1 (4.2)
	Unknown	24 (6.1)	2 (8.3)
	Hispanic	54 (13.7)	3 (12.0)
SES	Education		
	High School or Less	159 (39.2)	10 (40.0)
	More than High School	247 (60.8)	15 (60.0)
Clinical Phenotype	Diagnosis		
	IPAH/HPAH	182 (44.2)	11 (44.0)
	CTD-APAH	132 (32.0)	13 (52.0)
	Other APAH	98 (23.8)	1 (4.0)
	BMI, kg/m ²	28.5 (24.5, 33.6)	26.7 (21.6, 30.4)
	WHO functional classification		
	I	30 (7.9)	3 (12.0)
	II	132 (34.7)	3 (12.0)
	III	190 (50.0)	17 (68.0)
	IV	28 (7.4)	2 (8.0)
	Six-minute walk distance	340 (260, 427)	240 (138, 349)
	Baseline hemodynamics		
	RA, mmHg	9 (5, 13)	10 (7, 14)
	mPAP, mmHg	49 (37, 58)	49 (43, 63)
	CO, L/min	4.0 (3.4, 5.1)	3.2 (2.8, 3.8)
	CI, L/min/m ²	2.2 (1.8, 2.7)	1.9 (1.5, 2.3)
	PVR, Wood units	8.8 (5.7, 12.9)	12.8 (8.8, 17.1)
Supportive Care	Advanced directive	129 (31.2)	10 (40.0)
HRQOL	EmPHasis-10	26 (17, 34)	32 (22, 42)
	SF-12		
	Physical Health	34.7 (30.1, 38.2)	36.6 (31.1, 40.2)
	Mental Health	49.1 (42.1, 54.8)	41.5 (38.8, 50.2)
Treatment	Supplemental Oxygen	168 (40.8)	16 (64.0)
	PAH Targeted Therapy		
	Oral	352 (85.2)	22 (88.0)
	Inhaled	33 (8.0)	1 (4.0)
	Parenteral	89 (21.6)	12 (48.0)