Participation in pulmonary hypertension support group improves patient-reported health quality outcomes: a patient and caregiver survey

Paresh C. Giri[1] ⬤, Gizelle J. Stevens[1] ⬤, Jeanette Merrill-Henry[2], Udochukwu Oyoyo[3] and Vijay P. Balasubramanian[4]

[1]Pulmonary and Critical Care Department, Loma Linda University Medical Center, Loma Linda, CA, USA; [2]Respiratory Care Department, Loma Linda University Medical Center, Loma Linda, CA, USA; [3]Department of Dental Education Services, Loma Linda University School of Dentistry, Loma Linda, CA, USA; [4]Pulmonary and Critical Care Department, University of California San Francisco-Fresno, Fresno, CA, USA

Abstract

Support group participation has been shown to be effective in many chronic medical conditions. The evidence for integrating support group into pulmonary hypertension care and its effect on quality of life, physical and psychological well-being is limited. We sought to assess the effect of support group participation on quality of life in patients diagnosed with pulmonary hypertension and their caregivers. The emPHasis-10 questionnaire (a tool validated for quality of life assessment in pulmonary hypertension) was used to evaluate the effect of support group participation. Additional demographic and health-related quality measures were examined. Results showed that 165 subjects were enrolled in the study; 122 (74.4%) were patients with pulmonary hypertension, 41 (25.0%) were their caregivers, and 2 (0.02%) did not respond. The cohort was predominantly female ($n = 128$, 78%), Caucasian ($n = 10$, 61%), and the principal self-reported classification of pulmonary hypertension was World Health Organization Group 1 ($n = 85$, 51.8%) and the self-reported New York Heart Association Functional Class was II and III ($n = 43$, 57.3%). Most participants ($n = 118$, 71.5%) attended support groups and of them, a majority ($n = 107$, 90.6%) stated it helped them. There was no difference in quality of life as assessed by emPHasis-10 scores with support group participation (median score 30 vs 32, $p = 0.387$). There was self-reported improvement in understanding condition better including procedures such as right heart catheterization, medication compliance, and confidence in self-care ($p < 0.05$). Using multivariate logistic regression, baseline variables that were independently associated with emPHasis-10 scores for the entire cohort included knowledge of New York Heart Association Functional Class (odds ratio: 1.919, 95% CI: 1.004–3.67, $p = 0.04$) and greater distance traveled to visit pulmonary hypertension physician (odds ratio: 1.391, 95% CI: 0.998–1.94, $p = 0.05$). In conclusion, support group participation does not improve quality of life as assessed by emPHasis-10 scores but improves other meaningful health-related quality outcomes.

Introduction

In a chronic, life-threatening disease like Pulmonary Arterial Hypertension (PAH), the patients' perspective on their illness and its impact on themselves and their caregivers' quality of life (QOL), including symptomatic, psychosocial, intellectual, and spiritual dimensions of the disease, is of paramount importance.[1–3] Numerous studies unequivocally illustrate the emotional burden of PAH on patients manifested as depression,[4] anxiety,[5] stress,[6] feelings of isolation,[2] memory loss,[7] and lack of understanding of disease.[8] This emotional burden increases with worsening New York Heart Association Functional Class (NYHA-FC),[9] and could contribute to poor prognosis[10] and survival.[11]

Corresponding author:
Paresh C. Giri, 11234 Anderson Street, Loma Linda, CA 92354, USA.
Email: pgiri@llu.edu

A support group (SG) is described in the Merriam-Webster dictionary as: "a group of people with common experiences and concerns who provide emotional and moral support for one another". Participation in a SG may improve QOL. Positive salutary effects demonstrated in other chronic conditions include improvement in mood and perception of pain in breast cancer[12] and improved anxiety, depression, systolic blood pressure, heart rate, cholesterol, and reduced mortality in coronary artery disease.[13] There is a paucity of observational studies in the PAH cohort[14] and benefits of SG participation in patients with PAH have not yet been clearly elucidated.

Various QOL assessment surveys are available but only a few that are specific to PAH like the "emPHasis-10" questionnaire.[15,16]

The aim of this study was to determine if participation in a pulmonary hypertension (PH) SG has a significant effect on QOL in a population of patients with PH and their caregivers using the emPHasis-10 questionnaire.

Methods

Study population and design

We designed a questionnaire survey recruiting patients diagnosed with PH and their caregivers who attended the Pulmonary Hypertension Association's (PHAs) International Pulmonary Hypertension Conference and Scientific Sessions in Florida, USA on 29 June to 1 July 2018 (Phase I). Participants were also surveyed at the PH Clinic at our University Health Pulmonary Hypertension Regional Clinical Program from 1 February 2020 to 6 March 2020 (Phase II).

Participants were provided with a description of the study's purpose, expectations, confidentiality agreement and potential risk, and then gave verbal consent to participate in the survey. Investigators were stationed to either a designated research room at the PHA International Conference or a private room at the PH Clinic in our University to complete the study. Participants completed the 10–15 min survey online using Qualtrics XM® software.

Inclusion criteria were: age 18–80 years, a diagnosis of PH, any World Health Organization (WHO) Group, and the ability to read and understand English or Spanish. Exclusion criteria included patients unable to read English or Spanish. Participants needed to have no active mental illness such as psychosis that would affect their cognitive capabilities in completing the survey. To ensure diverse recruitment, a video describing the study was used to advertise prior to the commencement of the conference on the PHA webpage.

The study was approved by the Institutional Review Board at our University (IRB# 5180073). The study questionnaire was conducted on five patients as a pilot survey to enhance question development and interpretation of the patient responses. Patient response feedbacks were valued and survey adjusted accordingly.

Data collection

After obtaining their role as either patient or caregiver, data were collected to examine the participant's knowledge of their condition, treatment adherence, functional impact, and general socio-demographic information (age range, gender, racial background, and residence). Patients and their caregivers were quizzed on WHO group classification, NYHA-FC. Corroborating questions that described WHO group and NYHA-FC symptoms were asked to assess knowledge concordance. SG participation was assessed and perceived benefits were evaluated. Patients and their caregivers were queried about SG participation without providing an explicit definition of a SG. All participants were seated and given privacy to complete questionnaire but still within close vicinity to an investigator to address any issues. All questionnaires were conducted online with multiple-choice options, six-point Likert scale, and options to refuse any sensitive questions. The caregivers were asked to answer the questions on behalf of the patients, to the best of their ability.

Assessment of QOL

The primary outcome of the study was to assess differences in QOL with support group participation (SG(+)) using the emPHasis-10 questionnaire; a short 10-question survey each with a simple six-point (0–5) Likert scale validated for assessing health-related QOL specifically in patients with PH,[15] (survey in Supplemental Material). The emPHasis-10 questions assesses: (i) level of frustration due to breathlessness, (ii) breathlessness interrupting conversations, (iii) need to rest during the day, (iv) feelings of exhaustion, (v) energy level, (vi) ability to walk up flight of stairs without breathlessness, (vii) confidence in public, (viii) PH controlling their life, (ix) dependence, and (x) burden-like feelings. Scores range from 0 to 50 and higher scores indicate poorer QOL.[15]

Additional health-related secondary QOL measures were evaluated specifically in those that participated in SG(+), with six-point Likert scale questions. These assessed: (i) management of symptoms, (ii) understanding of condition and related procedures, (iii) access to emotional support, (iv) confidence in self-care, (v) pain management, (vi) medication cost awareness, (vii) lifestyle influences, and (viii) impact on healing.

Statistical analysis

Descriptive statistics were used to indicate the characteristics of the participants. Categorical variables were analyzed by frequency tables and expressed as counts and percentages. Continuous variables were analyzed by sample summary statistics and were expressed as median with

interquartile ranges and minimum/maximum. All analyses were imported from Qualtrics XM® and conducted using the Jamovi Software Version 1.2 (https://www.jamovi.org), Sydney, Australia. All tests of hypotheses were two-sided and conducted at an alpha level of 0.05. Missing data were not imputed and the number of participants with available data was reported. The emPHasis-10 scores were calculated according to the published instructions and was treated as continuous variable as well.[17] Mann–Whitney U test was used for independent sample test comparing SG(+) and no support group participation (SG(–)). The rest of the variables were compared between groups using Pearson X^2 test for equal proportions, or t- test for normally distributed data. Univariate and multivariate logistic regression analyses were used to determine the baseline characteristics associated with emPHasis-10 scores ≥ 30. An estimated sample size of at least 122 participants was needed for 80% power to detect a 25% difference in emPHasis-10 score.

Results

Patients

Of all the attendees at the PHA International Conference, 200 volunteered to participate in various research endeavors in the "Research Room", of which 130 (65.0%) enrolled in the questionnaire study. An additional 35 subjects from our University PH Clinic participated in the survey upon completion of their clinic visit. A total of 165 candidates were enrolled in the study and there were no withdrawal of participants and most of the pertinent questions were completed appropriately.

Baseline characteristics

A majority, $n = 122$ (74.4%) of the participants were patients diagnosed with PH, $n = 41$ (25.0%) were caregivers, and $n = 2$ (0.01%) did not respond to the question. This cohort was predominantly female, $n = 128$ (78.0%), Caucasian, $n = 101$ (61.0%), and the most prominent age group was 41–60 years ($n = 61$, 37.2%), Table 1. Due to the international nature of the conference, $n = 26$ (15.9%) resided outside the USA. The rest, $n = 139$ (84.1%) were from the US with patients from Southern USA comprising the highest majority, $n = 44$ (33.8%). The most common self-reported reason for PH was WHO Group 1, $n = 85$ (51.8%) and self-reported NYHA Functional class II and III, $n = 43$ (57.3%). The majority of patients were on PH-specific medications, the most common included Tadalafil, $n = 68$ (41.4%) and Ambrisentan, $n = 52$ (31.5%) and the median duration of PH diagnosis for entire cohort was five years.

Support group participation

A majority, $n = 118$ (71.5%), participated in SGs ($n = 109/130$, 83.8% in Phase I; 9/37, 24% in Phase II) and of them

most, $n = 107$ (90.6%), stated that SG participation helped them, Fig. 1.

The most common reasons to join a SG was to understand their condition better (42%) and to meet persons with similar condition (32%), Fig. 1. Many participants, $n = 69$ (42.1%) were involved in SGs affiliated to the PHA, Fig. 2.

Amongst the participants that did not want to join a SG ($n = 9$), "transportation issues" was the most (44%) common reason.

Median duration of PH diagnosis in SG(+) vs SG(–) was 5.75 vs 2.0 years, $p < 0.001$. More participants in SG(+) cohort (60.2 vs 29.8%, $p = 0.001$) had a self-reported diagnosis of WHO Group 1. Duration of SG participation was as follows: < 1 year: 20.2%, 1–3 years: 31.1%, 3–5 years: 11.8%, 5–10 years: 27.7%, more than 10 years: 9.2%. There was no difference in primary or secondary outcomes based on duration of SG(+). Table 1 outlines baseline characteristics differentiated by SG participation.

PH-specific QOL measures

There was no difference in emPHasis-10 scores with SG participation (median score 30 vs 32, $p = 0.387$), Table 2. No significant difference in any of the question domains of the emPHasis-10 survey was found when comparing SG(+) vs SG(–) cohorts. Similarly, there was no difference in emPHasis-10 scores when comparing Phase I (PHA Conference) vs Phase II (our University PH Clinic), median scores 29.5 vs 33, $p = 0.583$. In a univariate analysis of the entire cohort, four baseline variables significantly ($p < 0.15$) predicted emPHasis-10 scores: (i) knowledge of WHO Group, (ii) knowledge of NYHA-FC, (iii) greater distance traveled to visit PH physician, and (iv) gender, Table 3. Using multivariate logistic regression, baseline variables that remained independently associated with emPHasis-10 scores for the entire cohort included: (i) knowledge of NYHA-FC and (ii) greater distance traveled to visit PH physician. SG participation, duration of SG participation, and number of SG meetings participated in a year did not influence emPHasis-10 scores in this model. Most subjects (40.8%) with emPHasis-10 score < 30 traveled 5–20 miles to seek PH medical care, whereas most (35.6%) with scores $\geq$ 30 traveled > 50 miles, $p = 0.043$. A majority of participants, $n = 101$ (87.1%) who attended SG had a PH diagnosis ≥ 2 years as opposed to 59.5% in the SG(–) group, $p < 0.001$(data not shown). In the SG(+) cohort with PH diagnosis ≥ 2 years, frequency of emPHasis-10 scores above and below 30 was evenly distributed (42.1 vs 41.4%), but in the SG(–) cohort, scores favored higher than 30 (35.7 vs 23.8%, $p = $ ns).

Secondary health-related QOL outcomes

Owing to SG participation, > 80% of subjects stated there was improved: (i) self-healing, (ii) management of symptoms, (iii) adherence to medication, (iv) confidence in self-

Table 1. Patient and caregiver demographic and clinical characteristics based on support group participation.

Characteristics	Support group participation (n = 118)	No support group participation (n = 47)	p-Values[a]
Participant (n, %)			0.743
Patient	86 (74.1%)	36 (76.6%)	
Caregiver	30 (25.9%)	11 (23.4%)	
Age (n, %)			0.137
<20 years	2 (1.7%)	1 (2.2%)	
21–40 years	32 (27.1%)	6 (13.0%)	
41–60 years	47 (39.8%)	14 (30.4%)	
61–80 years	32 (27.1%)	22 (47.8%)	
> 81 years	2 (1.7%)	1 (2.2%)	
Unknown	3 (2.5%)	2 (4.3%)	
Gender (n, %)			0.378
Men	18 (15.3%)	9 (20.0%)	
Women	95 (80.5%)	33 (73.3%)	
Non binary	1 (0.8%)	2 (4.4%)	
Not reported	4 (3.4%)	1 (2.2%)	
Race (n, %)[b]			0.460
Caucasian	73 (61.9%)	28 (59.6%)	
African American	12 (10.2%)	5 (10.6%)	
Hispanic	11 (9.3%)	6 (12.8%)	
Other[c]	15 (12.7%)	2 (4.2%)	
Unknown	7 (5.9%)	6 (12.8%)	
Residence (n, %)			0.432
USA:	98 (76%)	40 (15.5%)	
West	11 (10.1%)	4 (20.0%)	
Mid-West	27 (24.8%)	3 (15.0%)	
South	34 (31.2%)	10 (50.0%)	
South-West	5 (4.6%)	0 (0%)	
North-East	21 (19.3%)	3 (15%)	
Outside USA[d]	6 (4.6%)	0 (0%)	
Unknown	3 (2.8%)	0 (0%)	
Patient perceived WHO Group classification (n, %)			0.001
Group I	71 (60.2%)	14 (29.8%)	
Group II	5 (4.2%)	3 (6.4%)	
Group III	8 (6.8%)	1 (2.1%)	
Group IV	6 (5.1%)	2 (4.3%)	
Group V	3 (2.5%)	1 (2.1%)	
Unknown	25 (21.2%)	26 (55.3%)	
Duration of condition			<0.001
Years, median (range)	5.75 (0.5–42)	2.0 (1–21)	
Affirmative understanding of term Functional Class[e]	64 (85.3%)	11 (14.6%)	<0.001
Patient-perceived NYHA-FC (n, %)[e]			0.164
FC I	10 (13.3%)	1 (1.3%)	
FC II	18 (24%)	2 (2.6%)	
FC III	21 (28%)	2 (2.6%)	
FC IV	6 (8%)	4 (5.3%)	
Unknown	9 (12%)	2 (2.6%)	
PH medication[f] (n, %)			
Sildenafil	29 (24.6%)	6 (12.8%)	0.094
Tadalafil	47 (39.8%)	21 (44.7%)	0.568
Riociguat	18 (15.3%)	11 (23.4%)	0.214
Ambrisentan	36 (30.5%)	16 (34.0%)	0.659
Bosentan	13 (11.0%)	5 (10.6%)	0.944
Treprostinil inhaled	12 (10.2%)	1 (2.1%)	0.084

(continued)

Table 1. Continued.

Characteristics	Support group participation (n = 118)	No support group participation (n = 47)	p-Values[a]
Treprostinil subcutaneous	26 (22%)	6 (12.8%)	0.174
Treprostinil oral	11 (9.3%)	3 (6.4%)	0.541
Epoprostenol intravenous	11 (9.3%)	2 (4.3%)	0.276
Macitentan	32 (27.1%)	13 (27.7%)	0.944
Iloprost inhaled	3 (2.5%)	2 (4.3%)	0.562
Selexipag	12 (10.2%)	6 (12.8%)	0.629
Digoxin	16 (13.6%)	7 (14.9%)	0.823
Calcium channel blockers	10 (8.5%)	7 (14.9%)	0.221
Other medications[g]	24 (20.3%)	3 (6.4%)	0.029

[a]p-Values obtained using Chi square test.
[b]Not exclusive.
[c]Other races included Native American, Asian, Middle eastern, Native Hawaiian.
[d]Outside of US include Puerto Rico and Australia.
[e]n = 75, the total number of respondents that answered "yes" to understanding of the term "Functional Class".
[f]Participants were asked to list all medications.
[g]Other medications: Lasix, spironolactone.
PH: pulmonary hypertension; WHO: World Health Organization; NYHA-FC: New York Heart Association Functional Class.
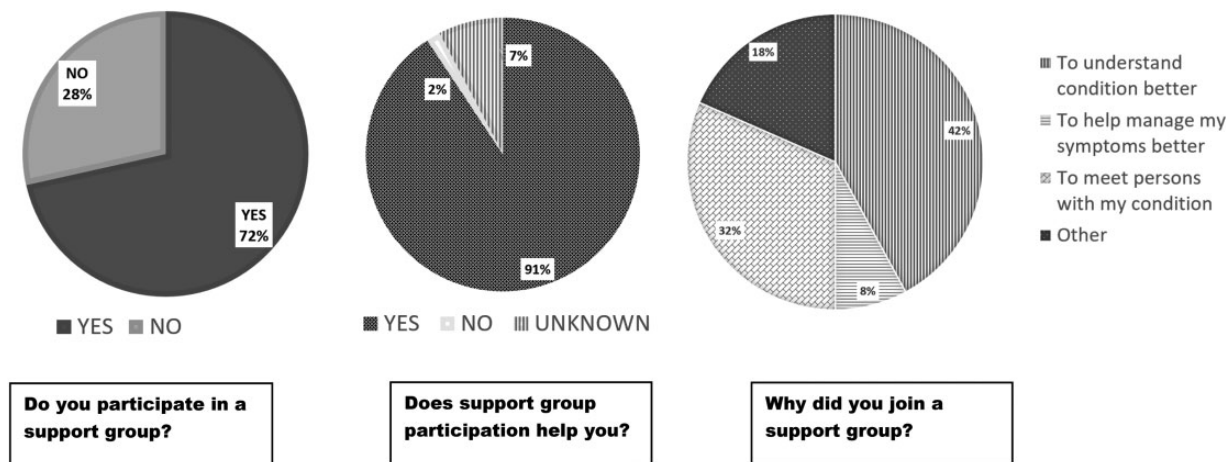

Fig. 1. Support group involvement and its perceived benefits.

care, (vi) understanding of the right heart catheterization procedure, and (viii) understanding of their condition, with significant differences in responses between patients and caregivers, Table 4.

SG participation did not affect: (i) adherence to fluid intake (45.1 vs 20.3%, $p = 0.212$), (ii) adherence to salt restriction (46.6 vs 17.8%, $p = 0.849$), or (iii) PH medication side effects (17 vs 7.9%, $p = 0.890$).

When comparing Phase I (PHA International Conference) to Phase II (our University PH Clinic) responses, the Phase I responders had higher number of caregiver participation (30.8 vs 2.7%, $p < 0.01$); higher SG participation (83.8 vs 24%, $p < 0.01$), higher Caucasians (73.8 vs 40.5%, $p = 0.008$); and a great proportion answered

that they knew the meaning of NYHA-FC (70 vs 5%, $p < 0.001$). There was no statistical difference in any other outcome with the exception that the Phase II participants were more likely to understand "how to exercise with PH" with median Likert scale 1 (Strongly Agree) vs 2 (Agree), p-value 0.023.

Parenteral prostacyclin use and SG

Of the entire cohort, $n = 59$ (35.8%) stated they use parenteral prostacyclin as part of their PAH therapy. More patients on parenteral prostacyclin vs oral PH therapy (41.5 vs 21.3%, $p = 0.014$) participated in SG. However, SG participation did not affect the emPHasis-10 scores

regardless of parenteral therapy. There was no difference in the secondary outcomes with the exception that patients on parenteral therapy were more likely to adhere to medications than those without parenteral therapy, Likert scale score 1 (Strongly Agree) in parenteral group vs score 2 (Agree) in non-parenteral group, $p = 0.026$. The distribution of emPHasis scores above or below 30 was similar in the (SG(+)) and SG(–) group for patients on parenteral prostacyclin therapy, Table 5.
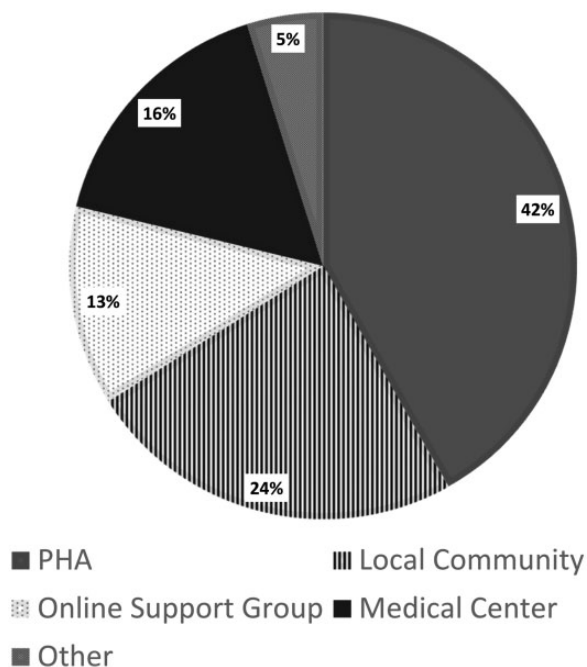

Fig. 2. Support group affiliation. Other responses included a combination of affiliations, Pulmonary Hypertension Camp, and Pulmonary Hypertension Association Australia.
PHA: Pulmonary Hypertension Association.

Understanding WHO group and Functional Class terminology

There was a discordance in the perceived knowledge of the WHO Group PH classification and NYHA Functional Classification when cross-referenced with corroborating questions that described WHO Group and NYHA-FC symptoms, in the entire cohort. SG participation did not improve discordance. Most participants had self-reported their Functional Class to be III, but the majority of their symptomology revealed Functional Class II. The sample size was too small to obtain statistical significance in the individual categories. However, in a multivariate analysis, knowledge of NYHA-FC remained predictive of emPHasis-10 scores, Table 3. Similarly, most participants self-reported their WHO Group to be 1, but chose other WHO groups in their answers to the corroborating questions. Sample size was too small to obtain statistical significance for individual WHO Groups.

Discussion

The purpose of this study was to survey the effects of SG(+) on the QOL in patients with PH and their caregivers. Central findings include the following: (i) a majority of those surveyed participated in SGs and found it helpful; (ii) benefits to SG participation included improvements in: feeling of self-healing, management of symptoms, adherence to medications, confidence in self-care, understanding of the right heart catheterization procedure and understanding of their condition; (iii) there was no difference in the primary outcome, QOL as assessed by emPHasis-10 scores, between SG(+) and SG(–) cohorts; and (iv) baseline variables associated with higher emPHasis-10 scores included perceived knowledge of NYHA-FC and distance traveled to visit their PH physician.

Table 2. The effects of support group participation on quality of life using emPHasis-10 questionnaire.

emPHasis-10 question (median score)	SG(+) (n = 118)	SG(–) (n = 47)	p-Values[a]
Frustration by breathlessness	4	4	0.775
Being breathless always interrupts my conversations	4	3	0.192
I always need to rest during the day	4	4	0.714
I always feel exhausted	4	4	0.996
I have no energy at all	4	5	0.343
When I walk up on flight of stairs I am very breathless	5	5	0.783
I am not confident at all in public places/crowds because of my PH	2	3	0.253
PH completely controls my life	3	3	0.296
I am completely dependent	2	2	0.307
I always feel like a burden	3	3	0.174
Total Score (Median)	30	32	0.387

[a]p-Values obtained using Mann–Whitney U.
SG(+): support group participation; SG(–): no support group participation; PH: pulmonary hypertension.
Note: emPHasis-10 score obtained from Yorke et al.[15]
Source: reproduced with permission from Yorke et al., 2014.[15]

Table 3. Unadjusted (univariate) and adjusted (multivariate) analyses for baseline variables that predict emPHasis-10 scores for entire cohort.

Variable	Unadjusted odds ratio (95% CI)	p-Values[a]	Adjusted odds ratio (95% CI)	p-Values
SG participation	1.261 (0.638–2.49)	0.504		
Duration of SG participation	0.967 (0.919–1.02)	0.199		
Number of SG meetings participated per year	1.119 (0.751–1.67)	0.581		
Knowledge of Primary WHO Group etiology	1.163 (1.012–1.34)	0.034	1.096 (0.942–1.28)	0.232
Knowledge of NYHA-FC	2.0 (1.073–3.73)	0.029	1.919 (1.004–3.67)	0.04
Distance traveled to visit PH physician	1.43 (1.042–1.97)	0.027	1.391 (0.998–1.94)	0.05
Gender (Female)	0.571 (0.310–1.05)	0.071	0.644 (0.345–1.20)	0.16
Age	1.02 (0.748–1.40)	0.881		
Parenteral prostacyclin therapy	0.892 (0.472–1.68)	0.724		

[a]p-Value of < 0.15 and < 0.05 considered significant in univariate and multivariate analyses, respectively.
SG: support group; NYHA-FC: New York Heart Association Functional Class; WHO: World Health Organization; PH: pulmonary hypertension.

Table 4. Analysis of secondary health-related outcomes with support group participation differentiated by patient and caregiver response.

Secondary outcome	Patient (n, %) (n = 87)	Caregiver (n, %) (n = 30)	p-Values[a]
Understand condition	75 (86.2%)	20 (66.7%)	0.008
Manage symptoms well	73 (83.9%)	18 (60%)	0.005
Adhere to medications	70 (80.4%)	14 (46.7%)	0.01
Adhere to diet	39 (44.8%)	10 (33.4%)	0.511
Receive emotional support	61 (70.1%)	21 (70%)	0.061
Understand oxygen management	61 (70.1%)	15 (50%)	0.005
Confidence in caring for self	77 (88.5%)	20 (66.7%)	0.02
Not feeling alone with the diagnosis	66 (75.9%)	19 (63.3%)	0.200
Better awareness of medication cost	69 (79.3%)	20 (66.6%)	0.014
Understand the right heart catheterization	77 (88.5%)	23 (76.6%)	0.001
Better pain management	45 (51.7%)	13 (43.3%)	0.439
Understand travel-related issue with the condition	60 (69%)	17 (56.6%)	0.091
Understand how to exercise with the condition	61 (70.1%)	14 (46.6%)	0.025
Help with healing	72 (82.7%)	22 (73.3%)	0.040

[a]p-values calculated by Chi-square.

Table 5. Differentiation of emPHasis-10 scores based on parenteral PH therapy and support group participation.

	Parenteral medication use[a]	SG(+) (n = 118) (n, %)	SG(−) (n = 47) (n, %)	p-Values[b]
	No	69 (58.5%)	37 (78.7%)	0.014
	Yes	49 (41.5%)	10 (21.3%)	
emPHasis-10 Score				
<30	No	32 (27.1%)	16 (34%)	0.058
	Yes	25 (21.2%)	4 (8.5%)	
≥30	No	37 (31.3%)	21 (44.7%)	0.118
	Yes	24 (20.3%)	6 (12.8%)	

[a]Parenteral medications include: intravenous epoprostenol, intravenous, subcutaneous or inhaled treprostinil, and inhaled iloprost.
[b]p-Values obtained using Chi square test.
SG(+): support group participation; SG(−): no support group participation.

SG participation and perceived benefits

A majority (71.5%) of the surveyed population participated in SGs. Participation was higher in the population attending the PHA Conference (83.8%) as opposed to that in the "real world" setting at our University PH Clinic (24%); a difference which could be attributed to the highly motivated nature of the cohort attending such an international conference. A majority (90.6%) stated that SG participation helped them in various categories including self-healing, management of symptoms, adherence to medications, confidence in self-care, understanding of the right heart catheterization procedure, and understanding of their condition. This could conceivably be a consequence of SG participation since topics discussed at SG sessions commonly address these issues (supplemental material, PHA webpage with topics discussed at SG meetings; https://phassociation.org/patients/living-with-ph/young-adults-with-ph/support-groups/) although it is impossible to ascertain an individual's exposure to any given topic. SG participation in other chronic conditions is consistent with improvement in these categories. A few examples include: improvement in healing from participating in a bereavement SG,[18] improved feelings of self-worth and self-value from a SG to prevent gender-based violence,[19] higher emotional, social, and physical functioning with a breast cancer SG involvement, and better understanding of medical condition from joining a hydrocephalus SG.[20] Nevertheless, the educational and emotional support value of any given SG is variable due to lack of a consistent curriculum and design and it is premature to draw conclusions of cause and effect without a uniformly structured SG and properly controlled studies. Comparisons to SG(–) for these secondary health-related QOL measures are unable to be performed as non-participators were not asked to answer how SG helped them.

There were a few differences in baseline characteristics when comparing the SG(+) and SG(–) cohort. First, the median duration of self-reported PH diagnosis was significantly longer in SG(+) (5.75 vs 2.0 years) and a majority (87.1%) in SG(+) had a PH diagnosis $\geq$ 2 years. This is consistent with data in other disease state SGs where participation is higher in patients with longer disease duration.[21] We surmise that longer duration of living with a disease exposes one to explore various additional coping mechanisms such as SG participation as proposed by the "THRIVE" model of coping.[22] Second, the self-reported diagnosis of WHO Group 1 was higher in the SG(+) indicating the class of patients that participate in PH SGs in this study. However, patient/caregiver understanding of WHO Group appears questionable as there was poor concordance between patient-reported and investigator-corroborated WHO Groups in both SG(+) and SG(–) cohorts.

The most common reasons to join a SG were to understand their condition better (42% responders) and to meet persons with similar condition (32% responders), a sentiment echoed in numerous surveys of patients with PAH and chronic thromboembolic pulmonary hypertension (CTEPH).[23,24] A majority (42.1%) were involved in SGs affiliated to the PHA which underscores the sample population and highlights the widespread SG opportunities made available to this cohort by the PHA. Amongst the participants that did not want to join a SG, "transportation issues" was selected by most (44%) as the reason. This emphasizes the social constraints to SG involvement.

More patients participated in SG if they were on parenteral prostacyclin and alternatively, more patients chose *not to* participate in SG if they were not on parenteral prostacyclin therapy which alludes to the knowledge and support-seeking behavior exemplified by patients on chronic, complex medical therapy[22] such as chronic parenteral prostacyclin.

SG participation and QOL assessed by emPHasis-10

To our knowledge, this is the first study in the United States to examine the effects of SG participation on patients with PH and their caregivers using the emPHasis-10 QOL questionnaire. We found no statistical significance in the emPHasis-10 scores with SG participation. There may be several plausible reasons. First, the majority of the subjects surveyed were attending an international conference, of which a few volunteered to participate in the various researches ongoing in the designated "Research Room" filtering the population to an extremely motivated cohort driven to learn about and participate in the care of their disease. Second, the emPHasis-10 tool may not be sensitive enough to elucidate differences with SG participation in this refined population. Third, QOL is improved with PAH therapy[25–29] and may be competing with SG for effect on QOL; a larger sample size may be thus required for SG effects to be elucidated. For these reasons, we reassessed the survey in a Phase II operation in a "real world" setting at our University PH clinic. Although, the sample population was small ($n = 37$), due to physical clinic visits being curtailed with COVID-19 restrictions, we did not find differences in QOL scores associated with SG participation. Since emPHasis-10 scores of > 30 differentiate patients with medically-determined (as opposed to patient-reported like in this study) NYHA-FC III and IV,[15] we chose to study differences in our population using this cutoff. There was no significant difference in distribution of emPHasis scores amongst SG(+) and SG(–) for the entire cohort (data not shown) as well as for those on parenteral prostacyclin therapy. When duration of PH diagnosis $\geq$ 2 years was included, we found that frequency of emPHasis-10 scores above and below 30 was evenly distributed in the SG(+) cohort (42.1 vs 41.4%), whereas in the SG(–) cohort, scores favored $\geq$ 30 (35.7 vs 23.8%, p = ns, data not shown) suggesting the potential benefit of SG participation in the group with longer disease duration. Using multivariate logistic regression, baseline variables that remained

independently associated with emPHasis-10 scores for the entire cohort included (i) knowledge of NYHA-FC and (ii) greater distance traveled to visit PH physician. To our knowledge, this is the first study to demonstrate these findings. Health literacy and education may,[30,31] or may not,[32] correlate with improved outcomes in heart failure and diabetes. Greater knowledge of COPD was associated with worse QOL[33] and could be due to higher levels of anxiety and depression associated with more knowledge.[34] These psychological symptoms are highly prevalent in the PH population[5] and further studies are required to assess the relationship of health literacy to QOL. Shorter distance to PH medical care may improve patient-perceived QOL perhaps, due to closeness to the medical team and the assurances thereof. The phenomenon of "distance decay",[35] which suggests lower healthcare facilitates usage in patients who live further away from medical care and its association with worse health-related outcomes is documented.[36] This has not been recognized in the PH population and further studies are warranted. Distance traveled to attend a SG meeting was not assessed in this survey and would be an interesting end point to analyze in future studies. Alternatively, it must be recognized that not all the domains of emPHasis-10 survey reflect the attributes that could potentially improve with SG participation and, as such, this tool may not be the apposite one to assess the effect of SG participation, even with above findings in the regression analyses. Questions regarding (i) "feeling like a burden", (ii) "PH controlling my life", and (iii) "having confidence in public places" are the likely areas assessed by the emPHasis-10 survey that SG participation *could* have a positive influence on; as is intrinsic to the definition of a SG (Merriam-Webster dictionary): "a group of people with common experiences and concerns who provide emotional and moral support for one another" and as is evident in the outcomes of involvement in SGs for various other chronic conditions.[12,13,20,37–40] Other (7 of 10) domains in the emPHasis-10 may not directly reflect on the benefits brought on by SG participation and further studies with complex QOL tools like CAMPHOR[16] or PAH-SYMPACT[41] may be required to elucidate differences.

Patient vs caregiver responses

This survey was unique in that both caregivers and the patients with PH were asked the same questions. The caregivers were asked to answer on behalf of the patient which inserts an inherent bias. The QOL questionnaire emPHasis-10 has not been validated in patient surrogates and differences can only be hypothesis-generating. Upon analysis of data, there were no significant differences in the responses except on secondary health-related outcomes outlined in Table 4. Most notably, unlike the caregivers, patients with PH thought: (i) participating in SG helped them understand their condition, (ii) manage symptoms well, (iii) adhere to medications, (iv) understand oxygen management better, (v) have confidence in caring for self, (vi) increased awareness of medication costs, (vii) improved understanding of RHC procedure, and (viii) improved healing. These fascinating differences between patient and caregiver perceptions can affect outcomes, e.g. in pain[42] and warrants further studies in PH.

Differences in phase I vs phase II responses

Phase I (PHA Conference) attendees had a higher SG participation rate compared to Phase II (our University PH Clinic), which is entirely conceivable given the "filtered" vs "real world" nature of the populations studied. More responders in Phase I than in Phase II acknowledged understanding the meaning of NYHA-FC, albeit their self-graded FC was not concordant with their self-reported symptoms. Remarkably, discordant answers notwithstanding the self-acknowledged familiarity with NYHA-FC were associated with a greater odds ratio (1.919) of achieving a higher emPHasis-10 score, as discussed above. Interestingly, inter-rater agreement on NYHA-FC even among physicians and nurses is poor[43] and a low concordance in patients and caregivers is not entirely unexpected.

In this era of shared decision making and patient engagement, understanding patient and caregiver perspectives as attempted by this study is key to cultivate holistic care. However, our study has several limitations. A sample of highly motivated group of patients and caregivers who traveled to the International PHA Conference may be inherently biased and may not be generalizable to the "real world". In an attempt to offset that bias, we surveyed patients and caregivers at our University PH Clinic and found substantive differences as reported. However, both populations studied were in essence, a "convenience sample" that was not randomized and the results can therefore be deemed to be hypothesis-generating rather than conclusive. The QOL questionnaire used for this study, emPHasis-10, is not validated for patient surrogates and may not be able to aptly capture subtle differences with SG participation and a more detailed questionnaire such as CAMPHOR[16] or PAH-SYMPACT[41] could be employed. However, both are relatively more complex and time-consuming to administer. There were instances when the patient and the caregiver of the same patient responded to the survey. Our survey questionnaire was not designed to capture that information accurately. However, we surmise that this is a small number. Recall bias may influence any survey-based study such as this one and enthusiasm for the results must be restrained. The educational and emotional support value of any given SG is variable due to lack of consistency in curriculum and design and it is premature to draw conclusions of cause and effect without uniformity in the SG structure and without properly controlled studies.

In conclusion, this is one of the first studies to demonstrate that PH SG(+) in patients with PH improves meaningful health-related outcomes including: self-healing, management of symptoms, adherence to medication,

confidence in self-care, understanding of the right heart catheterization procedure, and understanding of their condition. SG participation did not affect QOL as assessed by the emPHasis-10 questionnaire.

Author contributions

All authors made a substantial contribution to the concept or design of the work and analysis of data. All drafted the article or revised it critically for important intellectual content. All approved the version to be published. Each author as well has participated sufficiently in the work to take public responsibility for appropriate portions of the content.

Ethical approval

Loma Linda IRB approval #5180073.

Consent to participate

Verbal consent required per Loma Linda IRB recommendations.

Guarantor

Not applicable

Conflict of interest

The author(s) declare that there is no conflict of interest.

ORCID iDs

Paresh C. Giri https://orcid.org/0000-0002-0271-8572
Gizelle J. Stevens https://orcid.org/0000-0001-8323-2090

Supplemental material

Supplemental material for this article is available online.

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