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Physician perspectives on pulmonary hypertension in ILD: results of a cases-based survey

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Abstract

Background Pulmonary hypertension (PH) complicates the course of patients with interstitial lung disease (ILD) in 30–55% of cases and is associated with increased morbidity and mortality. The optimal timing of diagnostic right heart catheterization (RHC) and the impact of transthoracic echocardiographic (TTE) imaging on this decision remain uncertain. This study explores physician decision-making regarding PH suspicion in patients with ILD, and the necessity for TTE and RHC.

Methods A case-based survey was conducted among physicians from diverse geographic and professional backgrounds. Participants assessed anonymized ILD cases, providing their clinical suspicion of PH and recommendations for TTE and then RHC both before and after receiving TTE results. Predictive accuracy for PH was compared to the FORD index, a validated scoring system.

Results There were 10 cases provided, of which 3 had hemodynamically confirmed PH and 7 did not have PH. There were 42 respondents to the survey. Following the TTE results, the proportion of responses indicating high suspicion for PH increased in all PH cases and also increased in some non-PH cases. In PH cases, respondents accurately predicted mPAP ≥ 25 in 98.4% and PVR ≥ 3 Woods Units in 90.5% of responses, although only 24.6% and 20.6% matched the value ranges, respectively. In non-PH cases, mPAP < 25 and PVR < 3 Wood Units were correctly identified in 60.9% and 67.0% of responses, with 30% incorrectly predicting PH. Compared to the FORD index (sensitivity: 43.7%, specificity: 86.6%), respondents demonstrated higher sensitivity (88.9%) but lower specificity (70.1%) for PH diagnosis.

Conclusions Physicians demonstrate high sensitivity but moderate specificity in predicting PH, both with and without TTE results. The FORD index had greater specificity and may serve as a complementary tool, reducing the need for unnecessary RHCs. Standardized protocols are needed to facilitate detection of PH while optimizing the timing of RHCs in ILD patients.

Keywords Interstitial lung disease, Pulmonary hypertension, Survey

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Background

Pulmonary hypertension (PH) occurs during the clinical course of 30–55% of interstitial lung disease (ILD) patients [1], contributing to increased morbidity and mortality [2, 3]. Despite its devastating prognosis, the recent approval of inhaled treprostinil has underscored the growing importance of attaining a diagnosis of PH in a timely manner [4]. Clinical features, including symptoms, pulmonary function tests (PFT), the 6-minute walk test (6MWT), imaging studies such as computed tomography (CT), and blood tests like Brain Natriuretic Peptide (BNP)/N-terminal pro-BNP (NT-proBNP) are valuable for providing clues to the presence of PH in ILD patients [5]. Transthoracic echocardiography (TTE) is widely recognized as a key screening test, while right heart catheterization (RHC) is the gold standard for a definitive diagnosis [5, 6].

There is a wide inter-provider variation on the clinical index of suspicion for PH with many variables, including personal experience, weighing into the assessment of pretest likelihood of PH [6]. In addition, while there is a consensus on the necessity for TTE and RHC in the evaluation for PH in ILD patients, there is a paucity of research or guidelines on best practice and the impact of TTE results on the decision to perform RHC. Therefore, we conducted a physician survey using clinical information from 10 real-world cases to investigate the clinical gestalt of ILD providers in predicting PH, including if and when to proceed with a TTE, and how the results of TTE impact the decisions to proceed with RHC.

Methods

Survey protocols and study participants

We designed a survey that collected information on respondents' demographics, including nationality, practice setting, and their experience with patients. Additionally, we presented cases of actual ILD patients, including their brief clinical history, BNP or pro-BNP levels, PFT and 6MWT data, as well as chest CT images. Ten ILD cases were retrospectively selected from our institutional database based on the availability of comprehensive clinical data (PFT, TTE, and RHC) and their educational and clinical relevance, as determined through discussion among the co-authors. The proportion of cases with treatable PH approximated the estimated prevalence in a general ILD population. Respondents were asked to categorize their level of suspicion for PH as high, intermediate, or low and to indicate the necessity for TTE and RHC. Although there have been several definitions of PH, the respondents were asked their suspicion of PH based on the 5th World Symposium definition of precapillary PH; mean pulmonary arterial pressure (mPAP) ≥ 25 mmHg, pulmonary vascular resistance (PVR) ≥ 3 Wood Units, pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg [4]. While the 7th World Symposium definition is the current standard, the rationale for this older definition was based on the hemodynamic cut-offs used for approval of inhaled treprostinil for PH-ILD. The questionnaire used in the survey has been included as a supplementary file.

After providing TTE information in each case, respondents were again asked to categorize their level of suspicion for PH as high, intermediate, or low and to reassess the necessity for RHC as well as their estimate of the likely values for the mPAP and PVR. After each case response, the actual RHC values were presented. The survey was e-mailed to referring physicians to the Inova program as well as colleagues of the corresponding author. In the solicitations e-mail there was a link to a web-survey provider that hosted the online questionnaire (Survey Monkey® Inc., Palo Alto, CA, USA). Participation in this study was entirely voluntary, and no financial or other incentives were provided. Responses were collected between October 2024 and November 2024. Regarding the use of clinical data, all patient information was collected retrospectively, and the study was approved by the Institutional Review Board at Inova Fairfax hospital (IRB-2024-189). The invitation email for the survey clearly stated at the beginning that participation was voluntary and that only those who agreed to participate should proceed to complete the questionnaire. In accordance with national regulations and given the nature of the anonymous, minimal-risk survey, the IRB at Inova Fairfax hospital (IRB-2024-189) waived the requirement for written informed consent.

The performance characteristics of the respondents in predicting PH were compared to that of the FORD model, an objective scoring system. The FORD model, originally developed and validated in patients with idiopathic pulmonary fibrosis (IPF), incorporates four key components: the ratio of forced vital capacity (FVC) to diffusing capacity for carbon monoxide (DLco) (F), the oxygen saturation nadir during the 6MWT (O), race (R), and the distance ambulated during the 6MWT (D) [7]. The FORD index, a point scoring system for each variable, is provided in the Table 1.

Statistical analysis

After the web survey was completed, all data were initially entered into an Excel database, and the analysis was conducted using SPSS 23.0 and MEDCALC. 8.1.0.0. All values for continuous variables are expressed as median (interquartile range) and those for categorical variables are expressed as frequencies (%). Each respondent's response was expressed as a percentage of the total responses within each category. To evaluate the diagnostic performance of the respondents', the overall sensitivity and specificity of their responses were collated

Table 1 FORD index

Predictor	Category	Points
FVC%/ DLco%	FVC % predicted/ DLco % predicted	
	< 2	0
	2–3	2
	> 3	34
Oxygenation	Oxygen saturation (%) nadir during 6MWT	
	> 88	0
	83–88	19
	≤ 82	33
Race	White	0
	Non-white	20
Distance	Distance ambulated during 6MWT (m)	
	> 350	0
	250–350	9
	< 250	13

FVC: forced vital capacity; DLco: diffusing capacity for carbon monoxide; 6MWT: 6-minute walk test

and analyzed. Sensitivity was defined as the proportion of true positives correctly identified by the test, whereas specificity referred to the proportion of true negatives accurately identified. The sensitivity and specificity of the FORD index were also calculated based on established cutoff values [7].

Results

Demographics of participants

A total of 42 physicians participated in the survey, yielding a response rate of approximately 7% (42 out of an estimated 600 physicians contacted via email) and including 20 Asians, 17 Americans, and 5 Europeans (Fig. 1-A). Of the participants, 31 were pulmonologists, 2 were cardiologists, and the remainder were general practitioners. Their workplaces included 25 academic hospitals, 13 ILD-accredited centers, and 13 PH-accredited centers, 8 of which were dual-accredited for both ILD and PH.

The clinical experience of the participants is summarized in Fig. 1-B. The largest proportion of participants had over 20 years of experience treating ILD patients ($n = 14$, 33.3%), treated 10 to 50 ILD patients per year ($n = 19$, 45.2%), and treated fewer than 5 ILD-PH patients annually ($n = 17$, 40.5%).

Level of clinical suspicion for PH and the necessity of TTE and RHC evaluation

The clinical characteristics and test results of the 10 cases are presented in Table S1. There were 10 cases presented to the respondents, of whom 3 had hemodynamically confirmed PH (case #1, 5, 8), and 7 who did not have PH based on the definition employed. This information was blinded to the participants until they completed each case. The respondents index of PH suspicion, divided into high, intermediate, and low categories, is shown in Fig. 2, both before and after they were unblinded to the TTE results. For the PH cases, the proportion of responses indicating high suspicion increased following the TTE results (Case #1: 40.5–76.2%, Case #5: 28.6–81.0%, Case #8: 14.3–52.4%). However, even in some non-PH cases ($n = 3/7$), the proportion of responses indicating high suspicion increased following the TTE results (Case #3: 9.5–31.0%, Case #7: 4.8–21.4%, Case #10: 0 to 2.4%).

After the clinical information for each case was presented, a median of 95.2% responses (interquartile range: 93.5–97.6%) indicated the need for TTE. Responses regarding the necessity of RHC, both before and after the TTE information was presented, are shown in Fig. 3. Before obtaining TTE information, the proportion of responses indicating the necessity of RHC across the 10 cases had a median of 29.8% (IQR 19.1–33.3%). After the TTE information was provided, this proportion

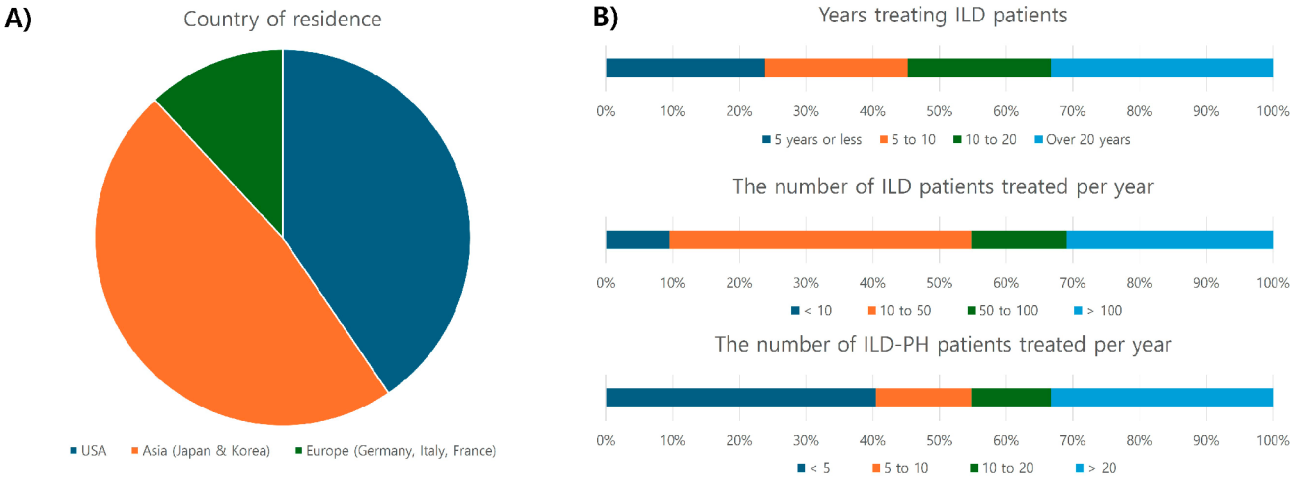

Fig. 1 Demographics for participants, **A**) country of residences, **B**) physician's experience

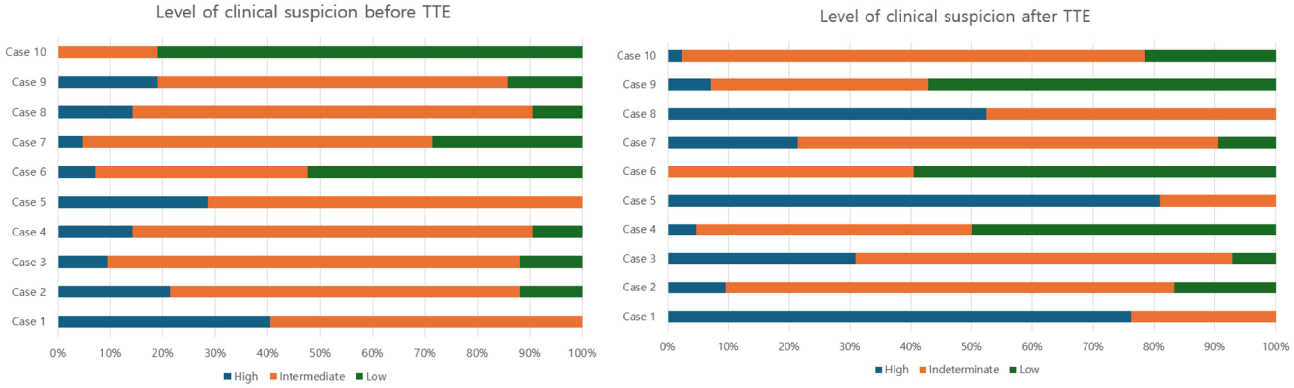

Fig. 2 Level of clinical suspicion for pulmonary hypertension, **A**) before echocardiography information, **B**) after echocardiography information, TTE: transthoracic echocardiography

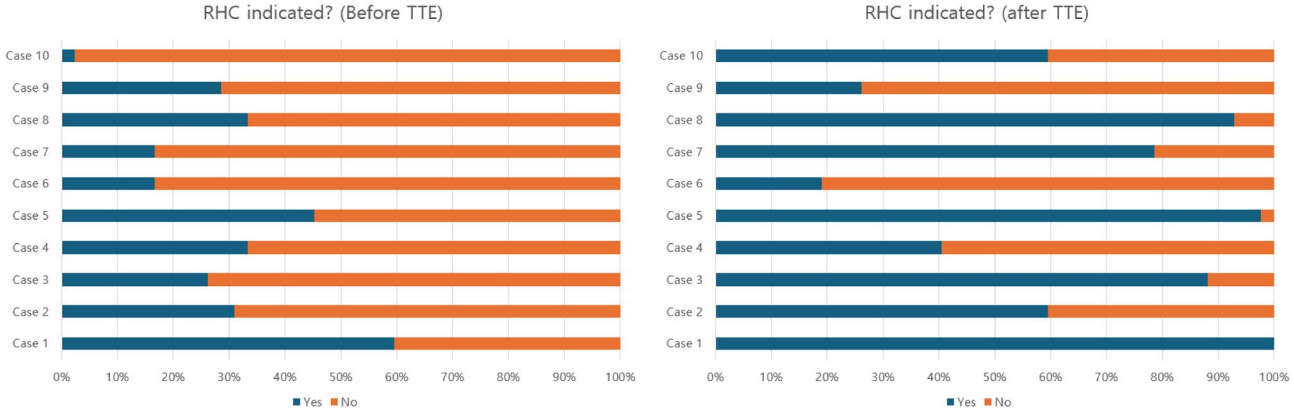

Fig. 3 Response on the necessity of right heart catheterization, RHC: right heart catheterization, TTE: transthoracic echocardiography

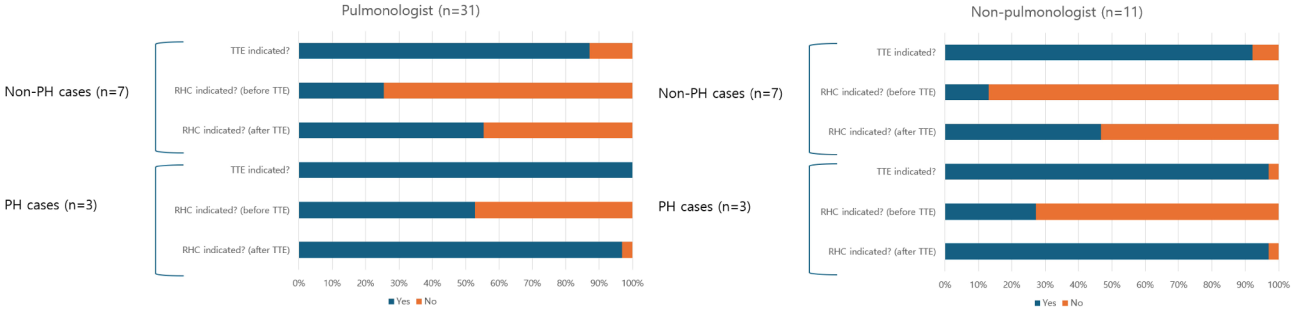

Fig. 4 Responses based on whether the participant was a pulmonologist or not

increased to a median of 69.0% (IQR 45.2–91.7%). In PH cases ($n=3$), the proportion of responses indicating the necessity of RHC before and after TTE information was 46.0% ($n=58/126$) and 96.8% ($n=122/126$), respectively. In non-PH cases ($n=7$), these proportions were 22.1% ($n=65/294$) and 53.1% ($n=156/294$), respectively (Figure S1).

Subgroup analysis based on the characteristics of participants

Responses from 31 pulmonologists and 11 non-pulmonologists were compared, and the results are presented in Fig. 4. Both pulmonologists and non-pulmonologists reported a higher necessity for RHC in cases with PH compared to those without PH. Compared to non-pulmonologists, pulmonologists indicated a greater need for RHC in both groups, with particularly higher responses suggesting the need for RHC before TTE information in cases with PH (52.7 vs. 27.3%). A subgroup analysis was

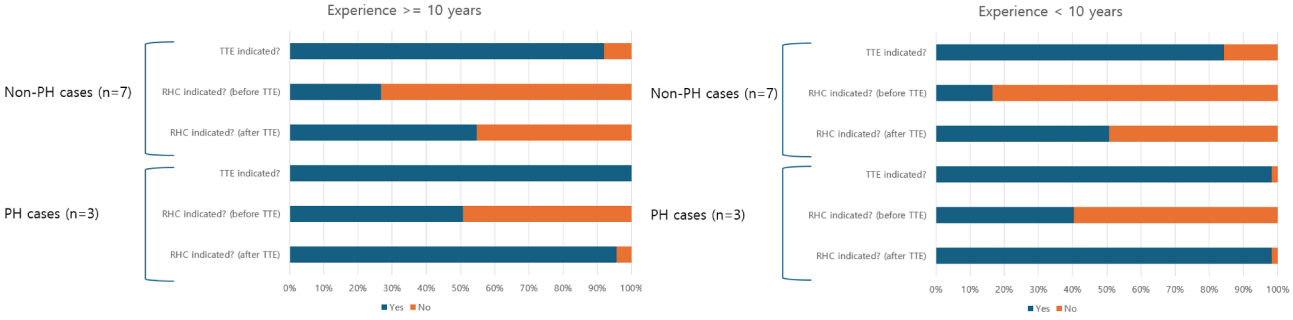

Fig. 5 Response according to the experience of participants

Table 2 Comparison of predicted and actual values of mPAP and PVR

PH cases (*n* = 3)					
mPAP	< 20 mmHg	21–25 mmHg	25–35 mmHg	35-45mmHg	> 45mmHg
#1 (actual: 48mmHg)	0	1 (2.4)	31 (73.8)	9 (21.4)	**1 (2.4)**
#5 (actual: 26mmHg)	0	0	**6 (14.3)**	23 (54.8)	13 (31.0)
#8 (actual: 30mmHg)	0	1 (2.4)	**24 (57.1)**	15 (35.7)	2 (4.8)
PVR	< 2 WU	2–3 WU	3–4 WU	4–5 WU	> 5 WU
#1 (actual: 9.0 WU)	0	5 (11.9)	21 (50.0)	12 (28.6)	**4 (9.5)**
#5 (actual: 3.5 WU)	0	0	**6 (14.3)**	14 (33.3)	22 (52.4)
#8 (actual: 3.3 WU)	0	7 (16.7)	**16 (38.1)**	15 (35.7)	4 (9.5)
Non-PH cases (*n* = 7)					
mPAP	≤ 20 mmHg	21–25 mmHg	25–35 mmHg	35-45mmHg	> 45mmHg
#2 (actual: 19mmHg)	**5 (11.9)**	12 (28.6)	20 (47.6)	5 (11.9)	0
#3 (actual: 20mmHg)	**3 (7.1)**	8 (19.1)	21 (50.0)	10 (23.8)	0
#4 (actual: 19mmHg)	**20 (47.6)**	14 (33.3)	5 (11.9)	3 (7.1)	0
#6 (actual: 22mmHg)	22 (52.4)	**17 (40.5)**	2 (4.8)	1 (2.4)	0
#7 (actual: 14mmHg)	**6 (14.3)**	14 (33.3)	16 (38.1)	6 (14.3)	0
#9 (actual: 20mmHg)	21 (50.0)	**14 (33.3)**	3 (7.1)	3 (7.1)	1 (2.4)
#10(actual: 17mmHg)	**14 (33.3)**	19 (45.2)	8 (19.1)	1 (2.4)	0
PVR	< 2 WU	2–3 WU	3–4 WU	4–5 WU	> 5 WU
#2 (actual: 2.0 WU)	6 (14.3)	**13 (31.0)**	14 (33.3)	6 (14.3)	3 (7.1)
#3 (actual: 2.0 WU)	3 (7.1)	**10 (23.8)**	17 (40.5)	8 (19.1)	4 (9.5)
#4 (actual: 1.2 WU)	**21 (50.0)**	13 (31.0)	4 (9.5)	4 (9.5)	0
#6 (actual: 2.4 WU)	27 (64.3)	**13 (31.0)**	1 (2.4)	0	1 (2.4)
#7 (actual: 1.0 WU)	**5 (11.9)**	15 (35.7)	17 (40.5)	3 (7.1)	2 (4.8)
#9 (actual: 1.8 WU)	**24 (57.1)**	12 (28.6)	2 (4.8)	3 (7.1)	1 (2.4)
#10 (actual: 2.6 WU)	17 (40.5)	**18 (42.9)**	7 (16.7)	0	0

Data are presented as number (percentage), mPAP: mean pulmonary artery pressure; PVR: pulmonary vascular resistance; WU: woods units

conducted based on the participants' experience, comparing the responses of participants with more than 10 years of experience ($n = 23$) to those with less than 10 years of experience ($n = 19$) (Fig. 5). In cases with PH, the proportion of responses indicating the necessity of RHC without knowledge of the TTE was numerically higher among participants with more than 10 years of experience (50.7%, $n = 35/69$) compared to those with less experience (40.4%, $n = 23/57$). However, responses regarding the necessity of TTE and RHC were similar overall in both groups. Subgroup analysis of responses based on the number of patients with ILD treated by participants also showed similar results (Figure S2).

Prediction of mPAP and PVR in PH cases

A comparison between the actual mPAP and PVR values and the predicted mPAP and PVR values provided by respondents in cases with PH is shown in Table 2. For mPAP, the overall proportion of cases in which the value range was accurately predicted for the three PH cases was 24.6% ((2.4 + 14.3 + 57.1)/3), while for the PVR it was 20.6% ((9.5 + 14.3 + 38.3)/3). An evaluation of the respondents ability to predict the presence of PH irrespective of the exact hemodynamic severity was then performed; of 126 (3 × 42) total responses in cases with PH, 124 (98.4%) accurately predicted mPAP ≥ 25 and 114 (90.5%) correctly predicted PVR ≥ 3 Wood Units. Only 14 responses

Table 3 Sensitivity and specificity analysis of PH using the FORD index

Case	FORD index	Sensitivity (%)	Specificity (%)	PPV (%)	NPV (%)
#1	48	42.6	92.5	38.5	93.6
#2	46	42.6	92.5	38.5	93.6
#3	50	42.6	92.5	38.5	93.6
#4	0	NA	NA	NA	NA
#5	86	12.8	99.3	66.7	91.1
#6	20	74.5	65.2	19.1	95.8
#7	28	61.7	73.6	20.6	94.6
#8	62	12.8	99.3	66.7	91.1
#9	34	59.6	77.6	22.8	96.6
#10	0	NA	NA	NA	NA

PH: pulmonary hypertension; PPV: positive predictive value; NPV: negative predictive value; NA: not available, FORD index is composed of the forced vital capacity (FVC)/diffusing capacity for carbon monoxide ratio (DLco) (F), oxygen saturation nadir during 6-minute walk test (6MWT) (O), race (R), and distance ambulated during 6MWT (D)

(11.1%) failed to predict the presence of PH by both criteria (mPAP ≥ 25 and PVR ≥ 3 Wood Units).

A comparison between the actual mPAP and PVR values and the predicted mPAP and PVR values provided by respondents in non-PH cases is also shown in Table 2. Of 294 total responses in those without PH, 179 (60.9%) accurately predicted mPAP < 25 and 197 (67.0%) correctly predicted PVR < 3 Wood Units. However, 88 responses (29.9%) incorrectly predicted the presence of PH by both criteria.

Sensitivity and specificity for PH diagnosis based on the FORD index and respondents' predictions

The FORD index for each case, along with the sensitivity and specificity based on the established cutoff values are shown in Table 3. Excluding the two cases with a FORD index of 0, for which sensitivity and specificity could not be calculated, the average sensitivity of the remaining 8 cases using the FORD index was 43.7%, while the average specificity was 86.6%. In these 8 cases, the sensitivity calculated using the respondents' predicted mPAP and PVR values was 88.9%, while the specificity was 70.1% (Table 3). In cases #4 and #10, where the FORD score was 0, the number of respondents who indicated the need for TTE was 40 (95.2%) and 22 (52.4%), respectively. The number of respondents who indicated the need for RHC was 14 (33.3%) and 1 (2.4%) before TTE information was provided, and 17 (40.5%) and 25 (59.5%) after TTE information was provided.

Discussion

Knowledge of the presence of PH in patients with fibrotic ILD has significant implications in the management of patients [8]. Not only does PH have significant prognostic implications [9], but there is increasing data that treating PH associated with ILD has important clinical benefits [4, 10, 11]. The diagnostic modality of choice remains RHC, but when to perform this in patients with ILD is uncertain. PH shares common symptoms with the underlying ILD and can "blend" in the background of increasing symptomatology rendering it more difficult to predict [12]. There are no good guidelines as to when to perform RHC in patients with ILD and practice habits vary widely in this regard [13, 14].

In our survey of 42 physicians across 3 continents we describe a spectrum of clinical suspicion that appears somewhat uniform across specialties and providers with varying expertise and geographic locations. There were subtle differences in responses based on years of experience, but there was otherwise no notable difference in predictive skills based on specialty, geographic location or other demographic features. In our study, while echocardiography uniformly increased the index of suspicion in those with PH, it also increased the index of suspicion in about half the patients who did not have PH.

Among the 42 respondents, RHC was deemed indicated in all the cases. Since 3/10 cases had PH, this translates to a % "hit rate" of 30% for detecting PH. There were no cases in which any of the respondents felt that RHC was not indicated. We are uncertain if this low threshold for obtaining a RHC reflects the participants' desire to know the numbers rather than truly reflecting what they would do in practice. A survey conducted among 55 clinicians in Europe reported that in cases where PH is suspected, echocardiography alone is performed on a median of 50% (IQR 20–73%) of patients, while echocardiography followed by RHC is performed on 35% (IQR 20–78%) of patients [13]. It is unknown what the "sweet spot" is in terms of the threshold to do RHC and what the optimal positivity rate for PH should be. Certainly if 90% of the RHCs demonstrate PH, then it is likely that many cases are being missed, while on the other end of the spectrum if only 10% have PH, then it is likely that this represents a too aggressive approach.

In terms of our respondents' predictive abilities, 11.1% (14/126) of responses for the 3 cases with PH were predicted to not have PH. On the other hand, for the patients without PH, 29.9% (88/294) were predicted to have PH. Several models for predicting ILD-PH have been proposed to date [15–17], among which the FORD model stands out as a simple tool that was specifically derived and validated in IPF [7]. Among the 8 cases in which a comparison between the sensitivity and specificity of the FORD index and the respondents' predictive ability could be made, it was observed that the respondents' predictions had higher sensitivity (88.9 vs. 43.7%) but lower specificity compared to the FORD index (59.5 vs. 70.1%). Once again, this likely reflects the respondents' assumption bias. While the clinician's overall accuracy in predicting PH in ILD patients seems better based on

the combination of sensitivity and specificity, they had the added advantage of knowledge of the echo results, whereas the FORD index does not incorporate echocardiographic parameters. Interestingly, the FORD index demonstrated a higher overall specificity of 86.6%, suggesting that it may be a reasonable tool for ruling out PH. Therefore, the FORD index might be helpful in contextualizing physicians' assumption that PH is present more often than it is and thereby reducing the need for RHCs.

There are several limitations to our current study. First, there are a number of biases that might have impacted the results of our survey. The fact that the respondents knew the survey was geared to the detection of PH might have resulted in an assumption or anticipatory bias. If participants knew that the cases were selected from patients who had received RHC, their pretest probability of PH might have been inherently elevated. In addition, specifically, the threshold to proceed with RHC might have been lowered so as not to miss any cases. However, we included only 3 cases of PH out of the 10 so at to replicate the estimate of PH in a general population of ILD patients [1]. Our response rate was rather low at about 7% (42 of an estimated 600 emails), and there might have been a responder bias. Given that approximately 40.5% of respondents manage fewer than 5 ILD-PH cases per year, this limited clinical experience may have influenced the survey results. Since individual identification of responders was not possible, repeated reminders could not be sent to increase survey participation rates. Furthermore, the 10 clinical cases were presented in a fixed sequence to all respondents, which may have introduced ordering effects. These could have influenced clinical judgment or diagnostic thresholds as the survey progressed. Our respondents performed well in their accuracy in predicting PH and likely reflected a group with interest and expertise in this area. The physicians responding to this survey were exuberant in their desire to perform RHC, but whether this actually reflects their clinical practice is uncertain. Additionally, although the FORD index was developed and validated in patients with IPF, our study included a broader spectrum of ILD subtypes. As the index has not been formerly validated in non-IPF ILD, however its performance characteristics in a broader ILD population appears to be similar [18]. A strength of our study lies in obtaining real-world data on the degree of PH suspicion and the use of diagnostic modalities from a diverse group of physicians, encompassing various specialties and providers with differing levels of expertise and geographic locations.

Conclusion

Our study demonstrated that physicians with interest and variable experience/expertise are good at predicting the presence of PH in ILD patients. Whether this level of expertise exists in a broader group of physicians is uncertain. The FORD model might be a useful complementary tool in risk-stratifying patients to undergo RHC. While RHC is necessary to diagnose PH in ILD patients, the timing of this is very important in optimizing the information and reducing the need for repeat RHCs in those ILD patients whose initial hemodynamics do not demonstrate "treatable PH".

Abbreviations

ILD	Interstitial Lung Disease
PH	Pulmonary Hypertension
TTE	Transthoracic Echocardiography
RHC	Right Heart Catheterization
mPAP	mean Pulmonary Arterial Pressure
PVR	Pulmonary Vascular Resistance
PCWP	Pulmonary Capillary Wedge Pressure
BNP	Brain Natriuretic Peptide
NT-proBNP	N-terminal pro-Brain Natriuretic Peptide
6MWT	6-Minute Walk Test
CT	Computed Tomography
PFT	Pulmonary Function Test
FVC	Forced Vital Capacity
DLco	Diffusing capacity for carbon monoxide
FORD	Forced vital capacity (FVC)/Oxygen saturation (O)/race (R)/distance (D) index
PPV	Positive Predictive Value
NPV	Negative Predictive Value
WU	Woods Units

Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s12890-025-03693-y.

Supplementary Material 1: The additional methods section and e-Figures are available online under "Supplementary Data."

Supplementary Material 2

Supplementary Material 3

Supplementary Material 4

Author contributions

SDN takes responsibility for the content of the manuscript, including the data and analysis. HCK and SDN contributed to the study conception and design. HCK, CSK, OS, CT, VK, OM, JW, SA, AS, AN and SDN contributed to the acquisition of data. HCK, CSK, OS, CT, VK, OM, JW, SA, AS, AN and SDN contributed to the analysis and interpretation of data. HCK and SDN drafted the manuscript. HCK, CSK, OS, CT, VK, OM, JW, SA, AS, AN and SDN had access to the final version of the manuscript.

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None.

Data availability

The datasets utilized and/or examined in the present study can be obtained from the corresponding author upon a reasonable request.

Declarations

Ethics approval and consent to participate

The study was approved by the Institutional Review Board at Inova Fairfax hospital (IRB-2024-189) and adhered to the principles outlined in the Declaration of Helsinki. Participation in this study was entirely voluntary, and no financial or other incentives were provided. The invitation email for the survey clearly stated at the beginning that participation was voluntary and

that only those who agreed to participate should proceed to complete the questionnaire. In accordance with national regulations and given the nature of the anonymous, minimal-risk survey, the IRB at Inova Fairfax hospital (IRB-2024-189) waived the requirement for written informed consent.

Consent for publication
Not applicable.

Competing interests
SDN is a consultant and is on the speaker's bureau for both Boehringer-Ingelheim and United Therapeutics. He has also received research funding from both Companies. He is also on the steering committee for IPF studies by Tvardi and Avalyn Pharma. CSK is a speaker for and has participated in advisory boards for United Therapeutics as well as Merck and Jannsen. CT has participated in advisory boards for Janssen. VK is a speaker for and has participated in advisory boards for Jannsen. OAS is on the speaker bureau for United Therapeutics, She is on the steering committee for PH studies for Gossamer and Insmed, and has also consulted for Merck, Janssen and Bayer. None of the other investigators have any conflicts of interest with regards to this study.

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References

1. Ang HL, Schulte M, Chan RK, Tan HH, Harrison A, Ryerson CJ, et al. Pulmonary hypertension in interstitial lung disease: A systematic review and Meta-Analysis. Chest. 2024;166:778–92.
2. Caminati A, Cassandro R, Harari S. Pulmonary hypertension in chronic interstitial lung diseases. Eur Respir Rev. 2013;22:292–301.
3. Gall H, Felix JF, Schneck FK, Milger K, Sommer N, Voswinckel R, et al. The Giessen pulmonary hypertension registry: survival in pulmonary hypertension subgroups. J Heart Lung Transpl. 2017;36:957–67.
4. Waxman A, Restrepo-Jaramillo R, Thenappan T, Ravichandran A, Engel P, Bajwa A, et al. Inhaled treprostinil in pulmonary hypertension due to interstitial lung disease. N Engl J Med. 2021;384:325–34.
5. Shlobin OA, Adir Y, Barbera JA, Cottin V, Harari S, Jutant EM, et al. Pulmonary hypertension associated with lung diseases. Eur Respir J. 2024. https://doi.org/10.1183/13993003.01200-2024.
6. Rahaghi FF, Kolaitis NA, Adegunsoye A, de Andrade JA, Flaherty KR, Lancaster LH, et al. Screening strategies for pulmonary hypertension in patients with interstitial lung disease: A multidisciplinary Delphi study. Chest. 2022;162:145–55.
7. Nathan SD, Chandel A, Wang Y, Xu J, Shao L, Watkins TR, et al. Derivation and validation of a noninvasive prediction tool to identify pulmonary hypertension in patients with IPF: evolution of the model FORD. J Heart Lung Transpl. 2024;43:547–53.
8. Ruffenach G, Hong J, Vaillancourt M, Medzikovic L, Eghbali M. Pulmonary hypertension secondary to pulmonary fibrosis: clinical data, histopathology and molecular insights. Respir Res. 2020;21:303.
9. Nikkho SM, Richter MJ, Shen E, Abman SH, Antoniou K, Chung J, et al. Clinical significance of pulmonary hypertension in interstitial lung disease: A consensus statement from the pulmonary vascular research institute's innovative drug development initiative-Group 3 pulmonary hypertension. Pulm Circ. 2022;12:e12127.
10. Waxman AB, Elia D, Adir Y, Humbert M, Harari S. Recent advances in the management of pulmonary hypertension with interstitial lung disease. Eur Respir Rev. 2022;31.
11. Dawes TJW, McCabe C, Dimopoulos K, Stewart I, Bax S, Harries C, et al. Phosphodiesterase 5 inhibitor treatment and survival in interstitial lung disease pulmonary hypertension: A bayesian retrospective observational cohort study. Respirology. 2023;28:262–72.
12. Olsson KM, Corte TJ, Kamp JC, Montani D, Nathan SD, Neubert L, et al. Pulmonary hypertension associated with lung disease: new insights into pathomechanisms, diagnosis, and management. Lancet Respir Med. 2023;11:820–35.
13. Montani D, Cifrián JM, Rojo RP, Nunes H, Meloni F, Ghio S et al. Unmet need in pulmonary hypertension-associated interstitial lung disease (PH-ILD): a clinician survey of real-world management of PH-ILD in Europe. ERJ Open Res. 2024;10.
14. Anderson JJ, Lau EM. Pulmonary hypertension definition, classification, and epidemiology in Asia. JACC Asia. 2022;2:538–46.
15. Parikh R, Konstantinidis I, O'Sullivan DM, Farber HW. Pulmonary hypertension in patients with interstitial lung disease: a tool for early detection. Pulm Circ. 2022;12:e12141.
16. Furukawa T, Kondoh Y, Taniguchi H, Yagi M, Matsuda T, Kimura T et al. A scoring system to predict the elevation of mean pulmonary arterial pressure in idiopathic pulmonary fibrosis. Eur Respir J. 2018;51.
17. Sobiecka M, Lewandowska K, Kober J, Franczuk M, Skoczylas A, Tomkowski W, et al. Can a new scoring system improve prediction of pulmonary hypertension in newly recognised interstitial lung diseases?? Lung. 2020;198:547–54.
18. Kim HC, Chandel A, King CS, Kim MJ, Shawabkeh M, Arunachalam A, et al. Performance of the FORD versus other available models for the noninvasive prediction of pulmonary hypertension in patients with interstitial lung disease. Lung. 2025;203:27.

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