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ORIGINAL ARTICLE

Pulmonary vascular stenosis scoring in fibrosing mediastinitis

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Abstract

Aims

This study aims to develop a scoring system for evaluating the degree of pulmonary vascular stenosis in fibrosing mediastinitis (FM).

Methods and results

A retrospective single-centre study was conducted on 56 patients with FM in China between April 2014 and August 2021. The involvement of pulmonary vessels in patients with FM was assessed using dual-phase computed tomography pulmonary angiography, and we found that 85.7% of the patients had both pulmonary artery (PA) and vein (PV) involvement. PA involvement was mainly located proximal to both the upper PA and the bilateral basal trunk levels in the lower lungs. The involvement of the superior PV was more common than that of the inferior PV, and the right inferior PV was the least involved. Most of these lesions exhibited moderate or severe stenosis. Additionally, a scoring system for evaluating the degree of pulmonary vascular stenosis was developed. A correlation analysis revealed a negative correlation between the final pulmonary vascular score and the pulmonary arterial pressure, pulmonary vascular resistance, and maximum tricuspid regurgitation velocity. The calculated score of 17.1 was the best cut-off value for the diagnosis of mild and severe pulmonary hypertension (PH).

Conclusion

We successfully developed a scoring system for pulmonary vascular stenosis that can be used to evaluate the severity of pulmonary vessel involvement and PH. This scoring system may be relevant in the future development of target-based strategies for percutaneous interventions.

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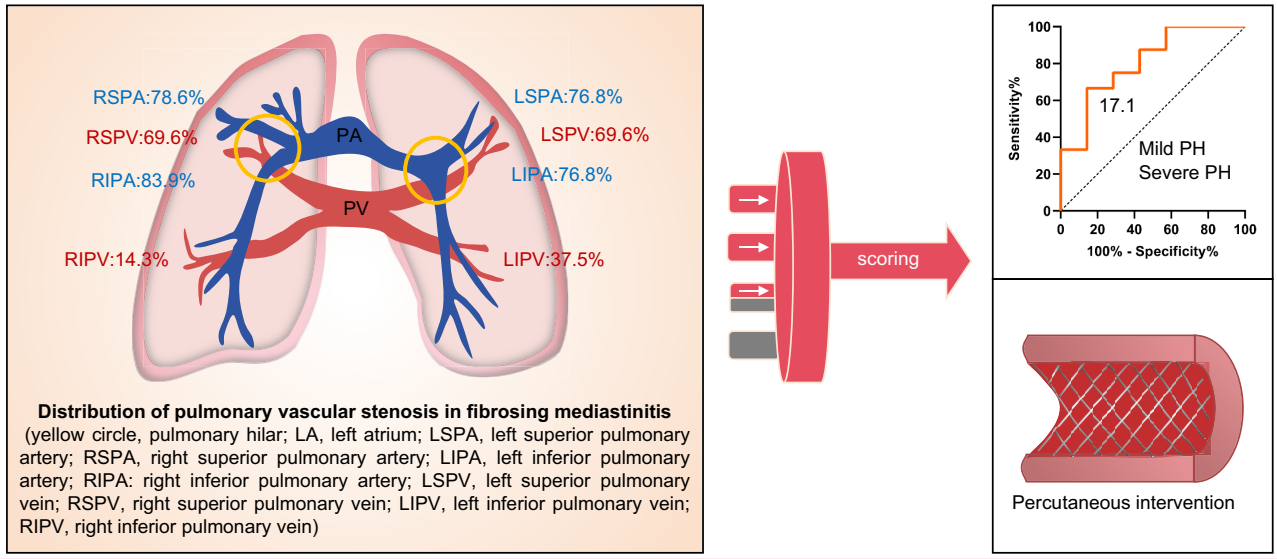
Graphical Abstract



Distribution of pulmonary vascular stenosis in fibrosing mediastinitis (yellow circle, pulmonary hilar; LA, left atrium; LSPA, left superior pulmonary artery; RSPA, right superior pulmonary artery; LIPA, left inferior pulmonary artery; RIPA: right inferior pulmonary artery; LSPV, left superior pulmonary vein; RSPV, right superior pulmonary vein; LIPV, left inferior pulmonary vein; RIPV, right inferior pulmonary vein)

Keywords fibrosing mediastinitis • pulmonary vascular stenosis • scoring system • pulmonary hypertension • severity • interventional therapy

Introduction

Fibrosing mediastinitis (FM) is characterized by proliferative fibrous tissue that replaces normal mediastinal fat, encasing, infiltrating, and compressing adjacent structures in the mediastinum, such as pulmonary vessels, bronchi, and the superior vena cava,[1–4] which can eventually progress to pulmonary hypertension (PH), right heart failure, and even death. The causes of FM vary regionally, with the most common being *Histoplasma capsulatum* infection in the USA and *Mycobacterium tuberculosis* infection in China.[5] Patients with FM are usually young in the USA, with most studies reporting a mean age of 20–40 years,[6,7] whereas the mean age of patients with FM was 69.5 years in the Chinese small-sample study.[8] Although histoplasma infection is common in the USA, the development of FM is typically rare. Wheat *et al.*[9] showed that only 3 of 100 000 patients with histoplasmosis infection developed FM. A previous study indicated that 11 of 31 patients with mediastinal granulomas had FM.[10] However, the incidence of FM caused by *M. tuberculosis* remains unknown.

PH associated with FM has a poor prognosis, with a 5-year survival rate of 56%.[11] Although the causes of FM-associated PH are multifactorial, the main causes are pulmonary artery stenosis (PAS) and pulmonary vein stenosis (PVS). Based on the involvement of pulmonary vessels in the mediastinum, FM can be divided into three subcategories: arterial, venous, and mixed.[4,12] Currently, there is no effective drug treatment for FM-associated PAS and PVS, and the surgical mortality rate is as high as 20%.[13] As an emerging technique, pulmonary vascular interventional therapy is the preferred treatment strategy for FM-associated PAS and PVS.[14–16] However, the formulation of interventional treatment strategies for pulmonary vascular stenosis associated with FM is complicated, and the pulmonary artery (PA), pulmonary vein (PV), and bronchus should be considered simultaneously. Therefore, understanding the characteristics and distribution of the PA and PV lesions in patients with FM is crucial for formulating interventional treatment strategies. In addition, the evaluation criteria of systemic vessels have long been used to evaluate the severity of the disease[17] and to judge whether intervention is necessary according to the degree of stenosis of a single pulmonary vessel. However, the PAs and PVs are a cohesive unit, and the degree of stenosis of a single pulmonary vessel does not reflect the degree of damage to the overall function of the pulmonary circulation.[18] Therefore, for patients with multiple branch stenosis of the pulmonary vessels, we need to establish a new evaluation system to assess the degree of pulmonary vessel damage. This new standard should serve as a guide for determining the necessity and specifics of further interventional treatments. Crucially, such an evaluation system must take into account the holistic functioning of both PAs and PVs.

Thus, we aimed to study the characteristics of pulmonary vessel involvement and develop a scoring system for evaluating the degree of pulmonary vascular stenosis using dual-phase computed tomography pulmonary angiography (DP-CTPA). This scoring system may be relevant in the future development of target-based strategies for percutaneous interventions.

Methods

This study was approved by the Ethics Committee of Gansu Provincial Hospital and conducted in accordance with the Declaration of Helsinki. The institutional review board waived the requirement for informed patient consent because of the retrospective nature of this study.

Patient selection

We retrospectively analysed patients diagnosed with FM who underwent CTPA between April 2014 and August 2021 at Gansu Provincial Hospital (Lanzhou, China). The diagnosis of FM was confirmed using contrast-enhanced chest computed tomography, and mediastinal malignancies were excluded.[19,20] Patients were excluded if the CTPA images were

unavailable. Mild PH was defined as a mean PA pressure (mPAP) of 21–24 mmHg with a pulmonary vascular resistance (PVR) ≥ 3 Wood units or an mPAP of 25–34 mmHg. Severe PH was characterized by an mPAP of ≥35 mmHg or an mPAP of ≥25 mmHg with a low cardiac index (<2.0 L/min m^2).[21]

Echocardiography

According to the current guidelines,[22] comprehensive transthoracic echocardiography was performed using a PHILIPS IE Elite machine with a 3.5 MHz transducer (Philips Medical System, Andover, MA, USA) by experienced certified sonographers prior to right heart catheterization (RHC). All values are presented as averages of three measurements. The parameters included the right ventricular area (RVA), right atrial area (RAA), tricuspid annual plane systolic excursion (TAPSE), tricuspid regurgitation (TR), and systolic PA pressure (sPAP; a detailed echocardiography protocol is given in the Supplementary data).[22,23]

CTPA imaging analysis and scoring

Three radiologists experienced with PH independently assessed all the chest DP-CTPA scans. Multiplanar reconstruction was used to examine arterial or vein (down to the subsegment level) narrowing, and grading was performed visually. Supplementary data and *Figure S1–5* show the pulmonary vascular terminology and stenosis measurements. The classification and assessment of the degree of pulmonary vessel stenosis were based on the classification criteria adopted from coronary artery stenosis in coronary computed tomography angiography (Coronary Artery Disease Reporting

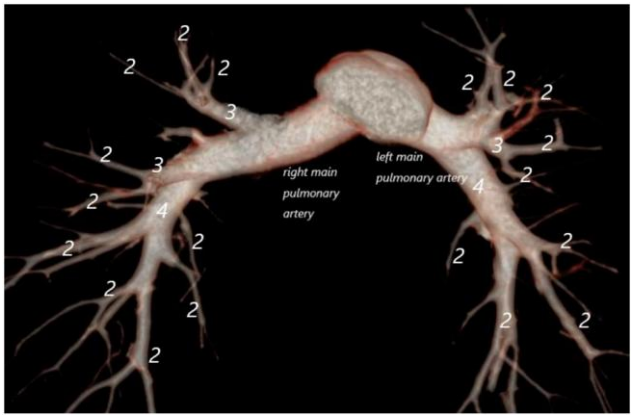


Figure 1 Pulmonary vascular tree.

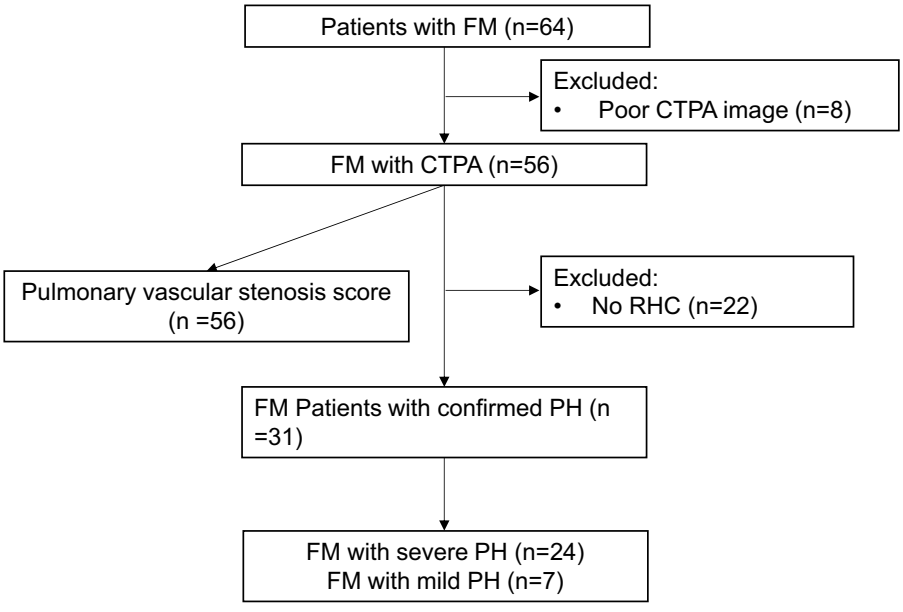


Figure 2 Flow chart of patient selection for the study. FM, fibrosing mediastinitis; PH, pulmonary hypertension; CTPA, computed tomography pulmonary angiography; RHC, right heart catheterization.

and Data System). This classification included categories of no stenosis (0%), minimal (1–24%), mild (25–49%), moderate (50–69%), severe (70–99%), and total occlusion (100%),[24] with corresponding values of 1, 0.99, 0.75, 0.5, 0.3, and 0. First, we used the diameter-defined Strahler system to score the pulmonary blood vessels.[25] The vascular order of the PA follows a hierarchical structure: segmental vessels are categorized as Order 2, forming a vessel of Order 3 when two Class 2 vessels converge. If one vessel of Order 3 intersects with two or more additional vessels of Order 2, it is classified as Order 4 (*Figure 1*). The vascular order of the PV is referred to as the PA. Second, the score for each vessel was obtained by multiplying the order by the value corresponding to the degree of stenosis. The total scores of segments PA and PV were calculated, and the PV score was normalized by the PA score; the lower score was chosen as the final score, considering the cohesive functioning of both the PA and the PV as a unit.

Statistical analysis

Continuous variables are presented as mean ± standard deviation or median [with interquartile range (IQR)]. Categorical variables are expressed as numbers (with percentages). Pearson's and Spearman's correlation coefficients were used to estimate the relationship between the final pulmonary vascular score and the parameters. Statistical significance was set at $P < 0.05$. The best cut-off value for the final pulmonary vascular score was obtained from the receiver operating characteristic (ROC) curve. This value represents the threshold that maximizes the classification accuracy of individuals with mild or severe PH, achieving greater sensitivity and specificity. The statistical software used included IBM SPSS 25, GraphPad Prism 8.0, and MedCalc 20.1.4.

Results

Study population

The screening and inclusion criteria for patients with FM are shown in *Figure 2*. A total of 56 patients with FM were enrolled, of whom 34 underwent RHC, which revealed that 31 had PH and 24 had severe PH.

Baseline characteristics of patients

In this study, 63% of the patients were female, and the mean age of the patients with a confirmed diagnosis of FM was 68.5 ± 9.0 years. The mean PA score was 19.6 ± 6.7; the median PV score was 19.8 (IQR 14.6–26.0), and the mean final pulmonary vascular score was 19.2 ± 6.7. Pleural effusion was observed in 15 (26.8%) patients, and 35 patients (62.5%) had a history of tuberculosis infection. The mean RVA in diastole was 26.7 ± 9.9 cm^2, and the mean RAA was 19.5 ± 6.8 cm^2; the mean estimated pulmonary arterial systolic pressure was 74.9 ± 23.0 mmHg, and the mean TR was 4.0 ± 0.8 m/s, as measured through echocardiography. Patients with FM had lower exercise capacity with decreased 6 min walking distance (6MWD: 269.8 ± 108.7 m), and 78.7% of patients were at WHO-FC II or III. Haemodynamic data demonstrated a mean mPAP of 39.8 ± 13.4 mmHg, a median PVR of 7.1 (IQR 5.1–9.8) Wood units, and a median PA wedge pressure of 5.5 (IQR 3.0–10.3) mmHg (*Table 1*).

Table 1 Baseline characteristics of patients

	FM ($n = 56$)
Age (years)	68.45 ± 9.00
Female sex	35 (62.50)
PA score	19.60 ± 6.86
PV score	19.75 (14.60–26.00)
Total pulmonary vascular score	19.22 ± 6.74
Clinical presentation	
Pleural effusion	15 (26.80)
Haemoptysis	6 (10.70)
Tuberculosis	
Confirmed	35 (62.50)
Suspected	20 (35.71)
No	1 (1.79)
Echocardiography	
TAPSE (mm)	19.42 ± 3.47 ($n = 26$)
TR (m/s)	3.96 ± 0.76 ($n = 44$)
RVA (cm^2)	26.72 ± 9.88 ($n = 25$)
RAA (cm^2)	19.49 ± 6.76 ($n = 25$)
Estimated PASP (mmHg)	74.89 ± 22.91 ($n = 44$)
Laboratory data	
ESR (mm/h)	13.50 (7.00–23.00) ($n = 24$)
NT-proBNP (ng/L)	617.50 (188.25–1729.25) ($n = 52$)
CRP (mg/L)	4.90 (2.00–13.35) ($n = 53$)
Exercise capacity	
6MWD (m)	269.82 ± 108.69 ($n = 22$)
WHO-FC I/II/III/IV	0/9/28/10 ($n = 47$)
Haemodynamics	
mPAP (mmHg)	39.82 ± 13.44 ($n = 34$)
sPAP (mmHg)	66.50 ± 23.40 ($n = 34$)
dPAP (mmHg)	24.00 (17.75–32.00) ($n = 34$)
PAWP (mmHg)	5.50 (3.00–10.25) ($n = 34$)
RAP (mmHg)	3.00 (2.00–4.00) ($n = 34$)
CI ($L/min/m^2$)	2.89 ± 0.95 ($n = 34$)
PVR (Wood units)	7.14 (5.06–9.82) ($n = 34$)
SvO_2 (%)	60.50 (54.75–64.00) ($n = 34$)

Data are presented as mean ± SD/median (range).
FM, fibrosing mediastinitis; TAPSE, tricuspid annual plane systolic excursion; TR, tricuspid regurgitation; RVA, right ventricular area; RAA, right atrial area; PASP, pulmonary arterial systolic pressure; NT-proBNP, *N*-terminal pro-brain natriuretic peptide; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; mPAP, mean pulmonary artery pressure; sPAP, systolic pulmonary artery pressure; dPAP, diastolic pulmonary artery pressure; PAWP, pulmonary artery wedge pressure; RAP, right atrial pressure; CI, cardiac index; PVR, pulmonary vascular resistance; SvO_2, mixed venous oxygen saturation; 6MWD, 6 min walking distance; WHO-FC, World Health Organization Functional Class; PA, pulmonary artery, PV, pulmonary vein.

Imaging characteristics of PA/PV involvement

Both the left and the right PAs were involved in 49 patients (87.5%); the right PA alone in 4 patients (7.1%); and the left PA alone in 3 patients (5.4%). A total of 48 (85.7%) patients had PVS, 34 (70.8%) had both left and right PV involvement, 7 (14.6%) had right PV involvement alone, and 7 (14.6%) had left PV involvement alone. Both PA and PV were involved in 48 patients (85.7%), PA alone in 8 patients (14.3%), and PV alone in 0 patients (0%; *Tables 2* and *3*).

The left upper lobe PA was involved in 43 patients (76.8%), mainly in the proximal segmental PA (58.9%), and the degree of stenosis was mostly moderate to severe (45.5–60.6%). The right upper lobe PA was involved in 44 patients (78.6%); 53.6–63.5% of the lesions were in the proximal segmental PA, and 46.7–65.7% of the lesions were completely occlusive. The left lingual artery was involved in 32 patients (57.1%), mainly at the common trunk level, and the degree of stenosis was mostly moderate to severe (50.0%). The right middle lobe PA was involved in 33 patients (58.9%), mainly at the common trunk level, and the degree of stenosis was mostly moderate to severe (45.5%). The left lower lobe PA was involved in 43 patients (76.8%), primarily at the basal trunk level (42.9%), and the degree of stenosis was mostly moderate to

Table 2 Number and percentage of patients with pulmonary vascular stenosis ($n = 56$)

Pulmonary vessel	No stenosis	Stenosis
PA		
LULA	39 (69.6)	17 (30.4)
LULA-1	23 (41.1)	33 (58.9)
LULA-2	23 (41.1)	33 (58.9)
LULA-3	24 (42.9)	32 (57.1)
LLPA	42 (75.0)	14 (25.0)
LLPA-1	38 (67.9)	18 (32.1)
LLPA-2	32 (57.1)	24 (42.9)
LLPA-3	44 (78.6)	12 (21.4)
LLPA-4	47 (83.9)	9 (16.1)
LLPA-5	46 (82.1)	10 (17.9)
RULA	38 (67.9)	18 (32.1)
RULA-1	34 (60.7)	22 (39.3)
RULA-2	26 (46.4)	30 (53.6)
RULA-3	21 (37.5)	35 (63.5)
RMLA	23 (41.1)	33 (58.9)
RLPA	38 (67.9)	18 (32.1)
RLPA-1	27 (48.2)	29 (51.8)
RLPA-2	26 (46.4)	30 (53.6)
RLPA-3	44 (78.6)	12 (21.4)
RLPA-4	56 (100.0)	0
RLPA-5	39 (69.6)	17 (30.4)
RLPA-6	55 (98.2)	1 (1.8)
PV		
LSPV	43 (76.8)	13 (23.2)
LSPV-1	24 (42.9)	32 (57.1)
LSPV-2	31 (55.4)	25 (44.6)
LSPV-3	32 (57.1)	24 (42.9)
LSPV-4	46 (82.1)	10 (17.9)
LIPV	46 (82.1)	10 (17.9)
LIPV-1	41 (73.2)	15 (26.8)
LIPV-2	51 (91.1)	5 (8.9)
RSPV	34 (60.7)	22 (39.3)
RSPV-1	36 (64.3)	20 (35.7)
RSPV-2	37 (66.1)	19 (33.9)
RSPV-3	32 (57.1)	24 (42.9)
RIPV	54 (96.4)	2 (3.6)
RIPV-1	50 (89.3)	6 (10.7)
RIPV-2	54 (96.4)	2 (3.6)

For information on the pulmonary arteries, veins, and their branches, see Supplementary data online, *Table S1*.

Table 3 Degree of stenosis of the pulmonary vessels

Pulmonary vessel	Minimal to mild	Moderate to severe	Occlusive
PA			
LULA ($n = 17$)	8 (47.06)	9 (52.94)	0
LULA-1 ($n = 33$)	7 (21.2)	20 (60.6)	6 (18.2)
LULA-2 ($n = 33$)	8 (24.2)	15 (45.5)	10 (30.3)
LULA-3 ($n = 32$)	6 (18.8)	16 (50.0)	10 (31.3)
LLPA ($n = 14$)	7 (50.0)	6 (42.9)	1 (7.14)
LLPA-1 ($n = 18$)	5 (27.8)	10 (55.6)	3 (16.7)
LLPA-2 ($n = 24$)	6 (25.0)	14 (58.3)	4 (16.7)
LLPA-3 ($n = 12$)	0	7 (58.3)	5 (41.7)
LLPA-4 ($n = 9$)	1 (11.1)	5 (55.6)	3 (33.3)
LLPA-5 ($n = 10$)	1 (10.0)	6 (60.0)	3 (30.0)
RULA ($n = 18$)	1 (5.56)	8 (44.4)	9 (50.0)
RULA-1 ($n = 22$)	2 (9.09)	9 (40.9)	11 (50.0)
RULA-2 ($n = 30$)	3 (10.0)	13 (43.3)	14 (46.7)
RULA-3 ($n = 35$)	3 (8.6)	9 (25.7)	23 (65.7)
RMLA ($n = 33$)	9 (27.3)	15 (45.5)	9 (27.3)
RLPA ($n = 18$)	8 (44.4)	10 (55.6)	0
RLPA-1 ($n = 29$)	0	18 (62.1)	11 (37.9)
RLPA-2 ($n = 30$)	9 (30.0)	20 (66.7)	1 (3.33)
RLPA-3 ($n = 12$)	1 (8.33)	4 (33.3)	7 (58.3)
RLPA-4 ($n = 0$)	0	0	0
RLPA-5 ($n = 17$)	1 (5.9)	2 (11.8)	14 (82.3)
RLPA-6 ($n = 1$)	0	1 (100.0)	0
PV			
LSPV ($n = 13$)	3 (23.1)	9 (69.2)	1 (7.7)
LSPV-1 ($n = 32$)	4 (12.5)	11 (34.4)	17 (53.1)
LSPV-2 ($n = 25$)	1 (4.0)	10 (40.0)	14 (56.0)
LSPV-3 ($n = 24$)	2 (8.3)	2 (8.3)	20 (83.3)
LSPV-4 ($n = 10$)	2 (20.0)	1 (10.0)	7 (70.0)
LIPV ($n = 10$)	2 (20.0)	6 (60.0)	2 (20.0)
LIPV-1 ($n = 15$)	4 (26.7)	8 (53.3)	3 (20.0)
LIPV-2 ($n = 5$)	1 (20.0)	3 (60.0)	1 (20.0)
RSPV ($n = 22$)	8 (36.4)	9 (40.9)	5 (22.7)
RSPV-1 ($n = 20$)	4 (20.0)	5 (25.0)	11 (55.0)
RSPV-2 ($n = 19$)	3 (15.8)	5 (26.3)	11 (57.9)
RSPV-3 ($n = 24$)	2 (8.3)	8 (33.3)	14 (58.3)
RIPV ($n = 2$)	1 (50.0)	0	1 (50.0)
RIPV-1 ($n = 6$)	2 (33.3)	3 (50.0)	1 (16.7)
RIPV-2 ($n = 2$)	0	1 (50.0)	1 (50.0)

For information on the pulmonary arteries, veins, and their branches, see Supplementary data online, *Table S1*.

severe (58.3%). The right lower lobe PA was involved in 47 patients (83.9%), mainly at the basal trunk level (53.6%), and the degree of stenosis was majorly moderate to severe (66.7%; *Tables 2* and *3*). The PAS of all lower lobes was located in the intra-pulmonary region (100%; *Figure 3A* and *B*), and the PAS of the upper lobe was rarely observed in the extra-pulmonary region (3–9%; *Figure 3C* and *D*).

A total of 39 patients had left superior PVS (69.6%); the stenosis was mainly located at the proximal first tributary of the PV (42.9–57.1%), and most lesions were found to be completely occluded (53.1–83.3%). Thirty-nine patients (69.6%) had a stenosis of the right superior PV (33.9–42.9%), and most of the lesions were completely occluded (55.0–58.3%). Although an inferior PVS is rare, 21 patients (37.5%) had a left inferior PVS and 8 (14.3%) had a right inferior PVS (*Tables 2* and *3*). The right superior PVS predominantly occurred in the intra-pericardial region (70%), with the narrowing primarily attributed to compression caused by a dilatation of the adjacent PA (79%; *Figure 4A* and *B*). The left superior PVS (79%) and left inferior PVS (83%) were mainly located in the extra-pericardial region (*Figure 4C* and *D*).

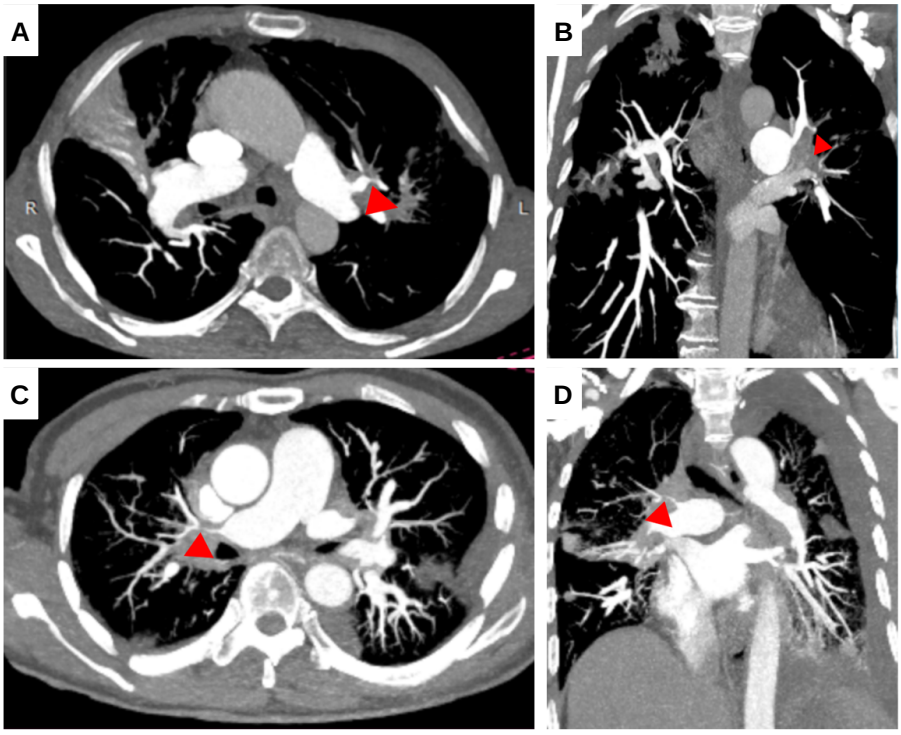


Figure 3 (*A* and *B*) Intra-pulmonary PA stenosis and (*C* and *D*) extra-pulmonary PA stenosis. Arrowheads: PA stenosis.

Correlation analysis between the final pulmonary vascular score and the clinical parameters

The final pulmonary vascular score was negatively correlated with sPAP ($r = -0.60$; $P < 0.0001$), diastolic PA pressure ($r = -0.42$, $P = 0.013$), mPAP ($r = -0.53$, $P = 0.001$), PVR ($r = -0.62$, $P < 0.0001$), or TR ($r = -0.46$, $P = 0.002$; *Figure 5*). The final pulmonary vascular score was weakly correlated with TAPSE ($r = -0.1$, $P = 0.641$), *N*-terminal pro-brain natriuretic peptide ($r = -0.14$, $P = 0.352$), 6MWD ($r = 0.02$, $P = 0.931$), and WHO-FC ($r = 0.18$, $P = 0.236$); however, there was no significant correlation (see Supplementary data online, *Table S2*).

Determination of cut-off values for the final pulmonary vascular score

The area under the ROC curve was 0.804 (95% confidence interval: 0.622–0.924), indicating a satisfactory overall performance of the final pulmonary vascular score in the diagnosis of severe PH. A calculated score of 17.1 was the best cut-off value for the diagnosis of mild and severe PH, with a sensitivity of 66.7% and a specificity of 85.7% (*Figure 6*).

Discussion

To the best of our knowledge, this study is the first to present a scoring system for assessing the degree of pulmonary vascular stenosis and to correlate the score with the clinical parameters. This study also identified the threshold for the final pulmonary vascular score to identify mild or severe PH.

Based on baseline characteristics, most of the patients enrolled in this study had combined PH and decreased exercise ability, which is consistent with those of previous studies.[4,26] In this study, RHC, the gold standard for haemodynamic diagnosis, confirmed the presence of significant haemodynamic impairment in patients with FM. The high incidence of pleural effusion in this cohort (26.8%) may be related to increased hydrostatic pressure secondary to PVS and right heart dysfunction secondary to PH.[4,11,27] Radiographic features (pulmonary and/or mediastinal) suggested a history of *M. tuberculosis* infection in most patients with FM in this study,[28] which supports previous findings from small-sample studies in China.[29]

Intriguingly, this study found that FM often involved the bilateral PAs and/or PVs, confirming the findings of previous small-sample-size studies in China.[4,8,29] However, studies conducted in Western countries have shown that unilateral involvement is more common.[2,7,30,31] The reason for this difference may be related to different triggers.[32] Tuberculosis-associated FM is usually associated with bilateral pulmonary vascular involvement, with PAs and PVs simultaneously involved, whereas histoplasmosis-associated FM may be associated with unilateral pulmonary vascular involvement, with less PV involvement.[33] Importantly, the present study showed that arterial stenosis was common in the segmental arteries of the upper lobe and in the common basal trunk arteries of the lower lobe. In addition, this study found that the majority of the narrowing observed in the right superior PV was attributed to compression resulting from the dilatation of the adjacent PA, and further investigation is required to determine the clinical significance of this phenomenon. In this study, the PAS of all lower lobes occurred in the intra-pulmonary regions (100%), and PAS of the upper lobe occurred in the extra-pulmonary regions (9%). The right superior PVS was mainly located in the intra-pericardium (70%), while the left superior PVS (79%) and the left inferior PVS (83%) were mainly located in the extra-pericardial regions; the right inferior PV was rarely involved.

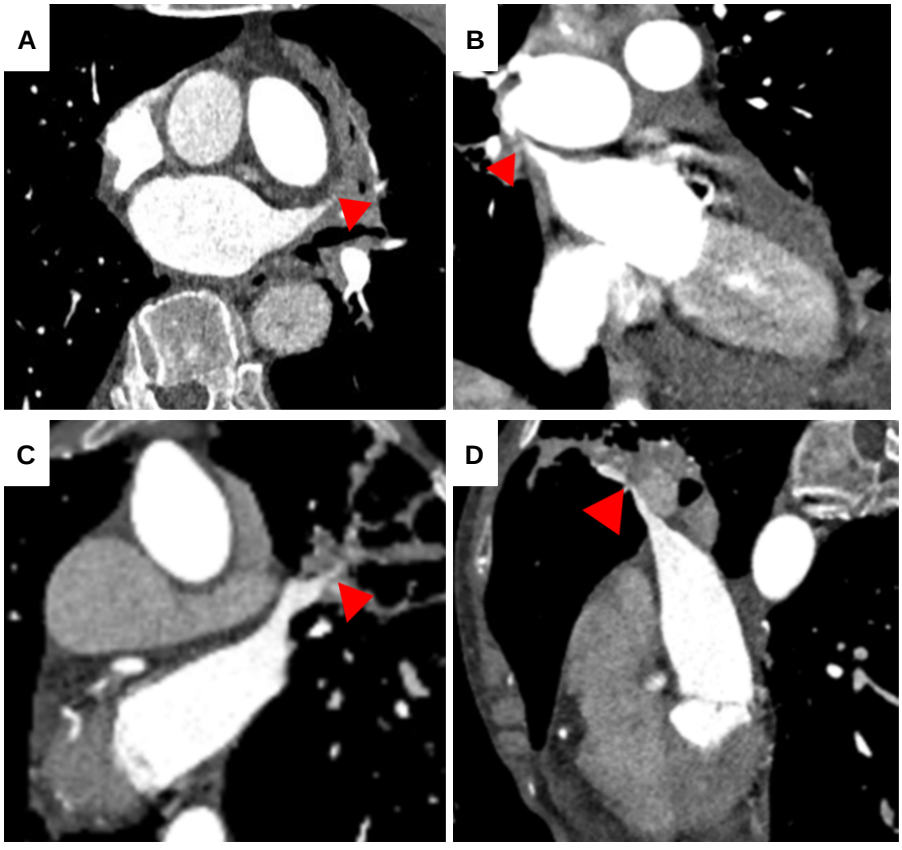


Figure 4 (*A* and *B*) Intra-pericardial PV stenosis and (*C* and *D*) extra-pericardial PV stenosis. Arrowheads: PV stenosis.

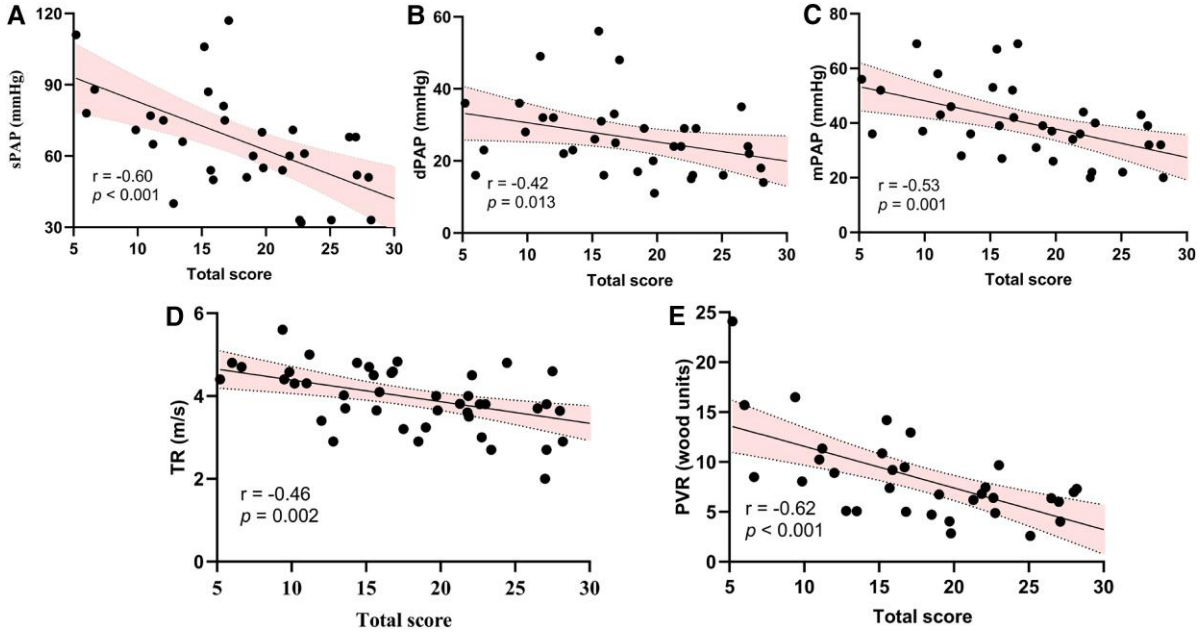


Figure 5 Correlation analysis between the total pulmonary vascular score and the clinical parameters. Scatterplot shows relationship between total pulmonary vascular score and (A) sPAP, (B) dPAP, (C) mPAP, (D) TR and (E) PVR. Solid circles denote data points, solid line represents least squares line of best fit, and dotted lines denote 95% confidence interval (CI). TR, tricuspid regurgitation; mPAP, mean pulmonary artery pressure; sPAP, systolic pulmonary artery pressure; dPAP, diastolic pulmonary artery pressure; PVR, pulmonary vascular resistance.

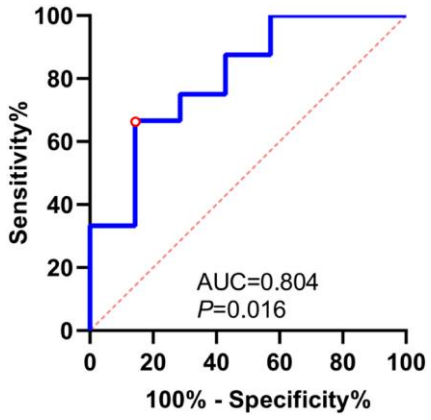


Figure 6 Receiver operating characteristic curves. AUC, area under the curve.

The distribution of involved vessels may be related to the hilar relationship between the PAs and the PVs. Percutaneous pulmonary vascular intervention is an emerging treatment for this disease, and detailed imaging information is essential for the development of rational interventional therapy strategies and the prevention of complications. For example, before interventional therapy, an assessment of the location of the affected vessel (intra- or extra-pericardial/pulmonary) may help to anticipate potential interventional complications and undertake preventive measures. For mixed FM, the general principle is to treat the PV first, followed by treatment of the PA. However, the restenosis rate of the PV is much higher than that of the PA, which greatly increases the difficulty of managing a mixed FM. Our study results revealed that although both PA and PV are involved in mixed FM, their distribution does not align on a one-to-one basis. Notably, the right inferior PV is rarely affected, presenting an opportunity for FM interventional therapy to prioritize treatment of the right inferior PA.

This study has developed a scoring system based on the degree of pulmonary vascular stenosis at the segmental level and can be used for quantitative assessment of this condition. The score decreases as the degree of stenosis increases, and a score of ≤17.1 suggests a high probability of severe PH. The scoring system was significantly correlated with PAP, PVR, and TR, where sPAP and PVR had a stronger negative correlation with the final score. This provides an important basis for a more accurate screening of patients requiring further invasive RHC. FM is a unique form of pulmonary vascular stenosis characterized by the coexistence of PAS and PVS. Therefore, this scoring system can be extended to other types of simple PAS and PVS in the future. Whether this score can provide a reference for the indication of intervention requires further investigation. However, other echocardiographic data reflecting right ventricular performance were not significantly correlated. In addition, there was no significant correlation among pulmonary vascular score, exercise capacity, and laboratory data, which may require a further study with a larger sample size.

Limitations

Our study also had limitations. Although FM is a rare disease, an imbalanced distribution of patients and the small sample size of the study still added bias to the concluding results. However, this pulmonary vascular scoring system has not yet been validated in other cohorts.

Conclusion

Pulmonary vascular stenosis caused by FM is mostly bilateral, and the PA and PV are involved simultaneously; the right inferior PV is rarely involved. We developed a scoring system to evaluate the overall degree of pulmonary vascular stenosis, which can also predict the PVR and sPAP. This scoring system may be relevant in the future development of target-based strategies for percutaneous interventions.

Supplementary data

Supplementary data are available at *European Heart Journal – Imaging Methods and Practice* online.

Consent

The institutional review board waived the requirement for informed patient consent because of the retrospective nature of this study.

Conflict of interest: None declared.

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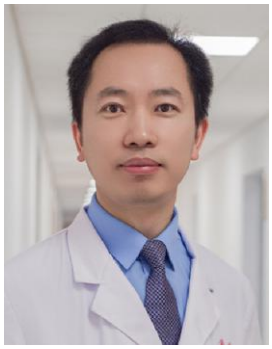
Data availab

The data underlying t e shared upon reasonable request from the corresponding author.

Lead author biography



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