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Relationship between High-Resolution Computer Tomography and FVC% Predicted for Classification of Pulmonary Hypertension in Systemic Sclerosis



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INTRODUCTION

- Pulmonary arterial hypertension (PAH) is one of the leading causes of mortality in scleroderma-spectrum disorders (SSc).
- It is defined as mean Pulmonary Artery Pressure (mPAP) > 20 mmHg, Peripheral Vascular Resistance (PVR) \geq 3 WU, and no/mild lung disease, as proposed by the 6th World Symposium on Pulmonary Hypertension (WSPH).
- An FVC threshold of 70% has been used as an adequate prognostic substitute¹ in different clinical trials to differentiate Group 1 (PAH) vs. Group 3 (PH due to chronic lung disease).

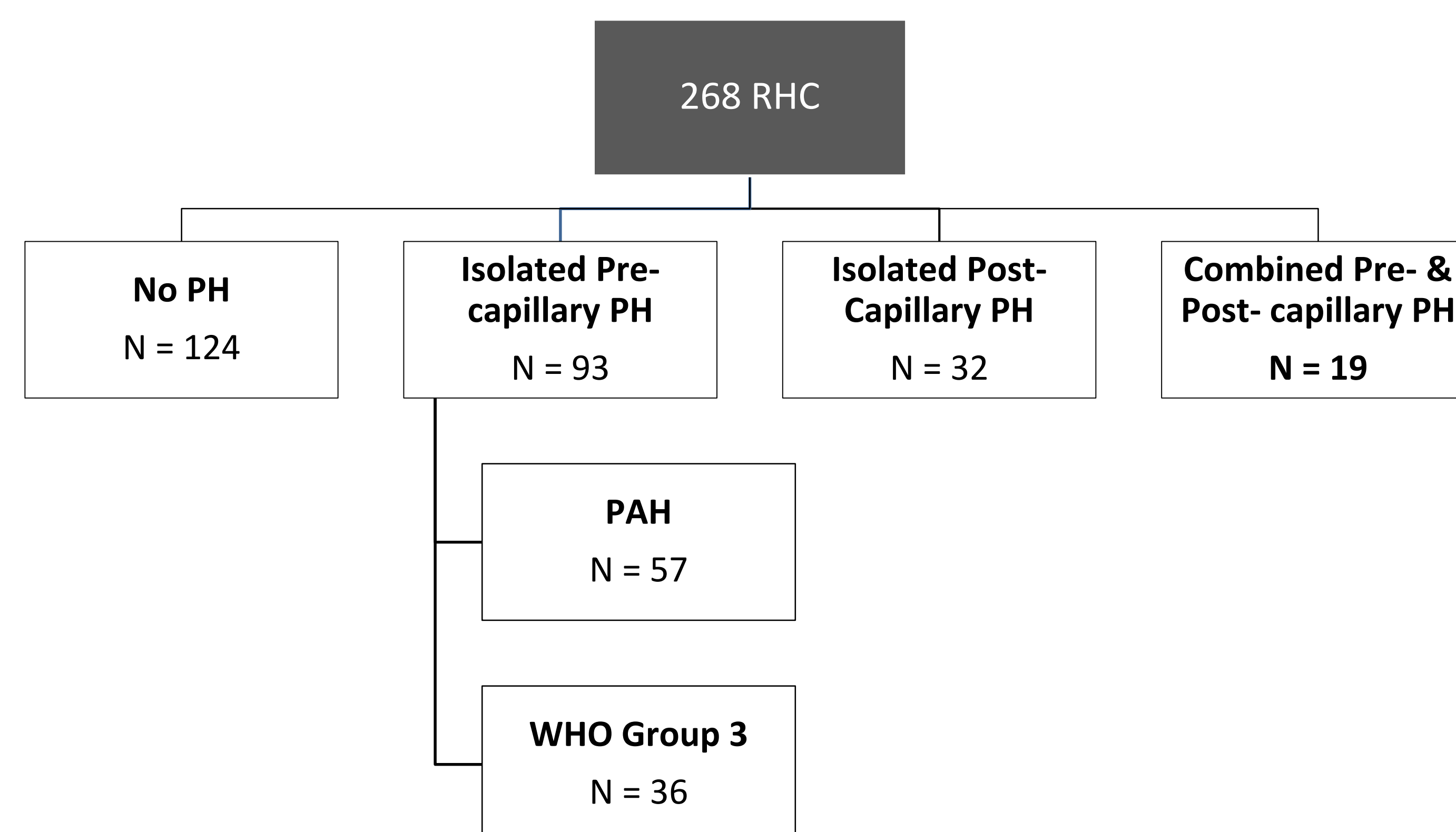
OBJECTIVES

- The objective of the current analysis was to assess the relationship between the degree of lung disease on high-resolution computed tomography (HRCT, gold standard) vs. FVC% in identifying those with PH due to Group 1 vs. Group 3 PH.

METHODS

- In this single center retrospective analysis, 268 patients with SSc who had right heart catheterizations (RHCs) at a tertiary center in US were included.
- HRCTs were reviewed by 2 thoracic radiologists who assessed the degree of total lung involvement in increments of 10% up to 30% or >30% lung involvement, and if there was concomitant emphysema.

Figure 1 : Classification according to the new Hemodynamic Definition of PH



METHODS continued

- If emphysema was present, it was classified as mild, moderate, or severe.
- Chronic lung disease was defined as HRCT showing >20% total lung involvement due to chronic lung disease; or if the total lung involvement due to chronic lung disease was 10-20% but the patient had concomitant moderate-to-severe emphysema.
- Each HRCT was categorized based on FVC% performed closest to the RHC into <70% or \geq 70%.
- Of the 268 RHCs, 57 had Group 1 and 36 had Group 3 PH based on the updated hemodynamic definition of PH, as proposed by the 6th WSPH (Figure 1)^{2,3}.
- In 75 of 93 patients with Group 1 or 3 PH, we had available HRCT data and 54 were reviewed by thoracic radiologists.
- Of 75 HRCTs, 34 (45%) patients had moderate-to-severe lung disease (based on the definition above), 20 had mild disease, and 21 did not have any chronic lung disease and/or significant emphysema.

RESULTS

Table 1: Relationship between HRCTs and FVC% in the Whole Cohort (N = 75)

FVC %, N(%)	HRCT consistent with moderate to severe chronic lung disease (N = 34)	HRCT - normal or mild chronic lung disease (N = 41)
FVC < 70%, 51 (68%)	25 (74%)	26 (63%)
FVC \geq 70 %, 24 (32%)	9 (26%)	15 (37%)
	Sensitivity 74% 95% CI (56%, 87%)	Specificity 37% 95% CI (22%, 53%)

Table 2: Relationship between HRCTs and FVC% in those with Chronic Lung Disease involvement on HRCT (N = 54)

a. FVC cut-off of < 60% vs. \geq 60%

FVC %, N(%)	HRCT consistent with moderate to severe chronic lung disease (N = 34)	HRCT - mild chronic lung disease (N = 20)
FVC < 60%, 21 (39%)	15 (44%)	6 (30%)
FVC \geq 60 %, 33 (61%)	19 (56%)	14 (70%)
	Sensitivity 44% 95% CI (27%, 62%)	Specificity 70% 95% CI (46%, 88%)

b. FVC cut-off of < 70% vs. \geq 70%

FVC %, N(%)	HRCT consistent with moderate to severe chronic lung disease (N = 34)	HRCT - mild chronic lung disease (N = 20)
FVC < 70%, 35 (65%)	25 (74%)	10 (50%)
FVC \geq 70 %, 19 (35%)	9 (26%)	10 (50%)
	Sensitivity 74% 95% CI (56%, 87%)	Specificity 50% 95% CI (27%, 73%)

c. FVC cut-off of < 80% vs. \geq 80%

FVC %, N(%)	HRCT consistent with moderate to severe chronic lung disease (N = 34)	HRCT - mild chronic lung disease (N = 20)
FVC < 80%, 47 (87%)	32 (94%)	15 (75%)
FVC \geq 80 %, 7 (13%)	2 (6%)	5 (25%)
	Sensitivity 94% 95% CI (80%, 99%)	Specificity 25% 95% CI (9%, 49%)

CONCLUSION

- FVC% misclassifies a large number of patients into Group 1 vs. Group 3 PH as it may be influenced by other SSc-related disease processes.
- FVC% can be influenced by the technique of the procedure, diaphragmatic involvement, and thoracic skin involvement of the disease.
- Future studies should incorporate the degree of HRCT involvement to differentiate between the 2 groups.

REFERENCES

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