

SUPPLEMENTAL MATERIAL

Online Supplement 1. Pulmonary Function Parameters

Cohort	SScPAH (n=166)	SSc-ILD-PH (n=49)	P value
FEV1	1.91 ± 0.60 (n=144)	1.47 ± 0.44 (n=45)	<0.001
FEV1 (% predicted)	78 ± 18 (n=129)	53 ± 13 (n=46)	<0.001
FVC	2.52 ± 0.82 (n=145)	1.75 ± 0.58 (n=45)	<0.001
FVC (% predicted)	80 ± 18 (n=131)	50 ± 11	<0.001
FEV1/FVC	76.4 ± 7.8 (n=144)	84.9 ± 8.3 (n=46)	<0.001
DLCO	10.5 ± 4.0 (n=122)	8.2 ± 4.2 (n=40)	<0.001
DLCO (% predicted)	52 ± 18 (n=108)	34 ± 14 (n=43)	<0.001

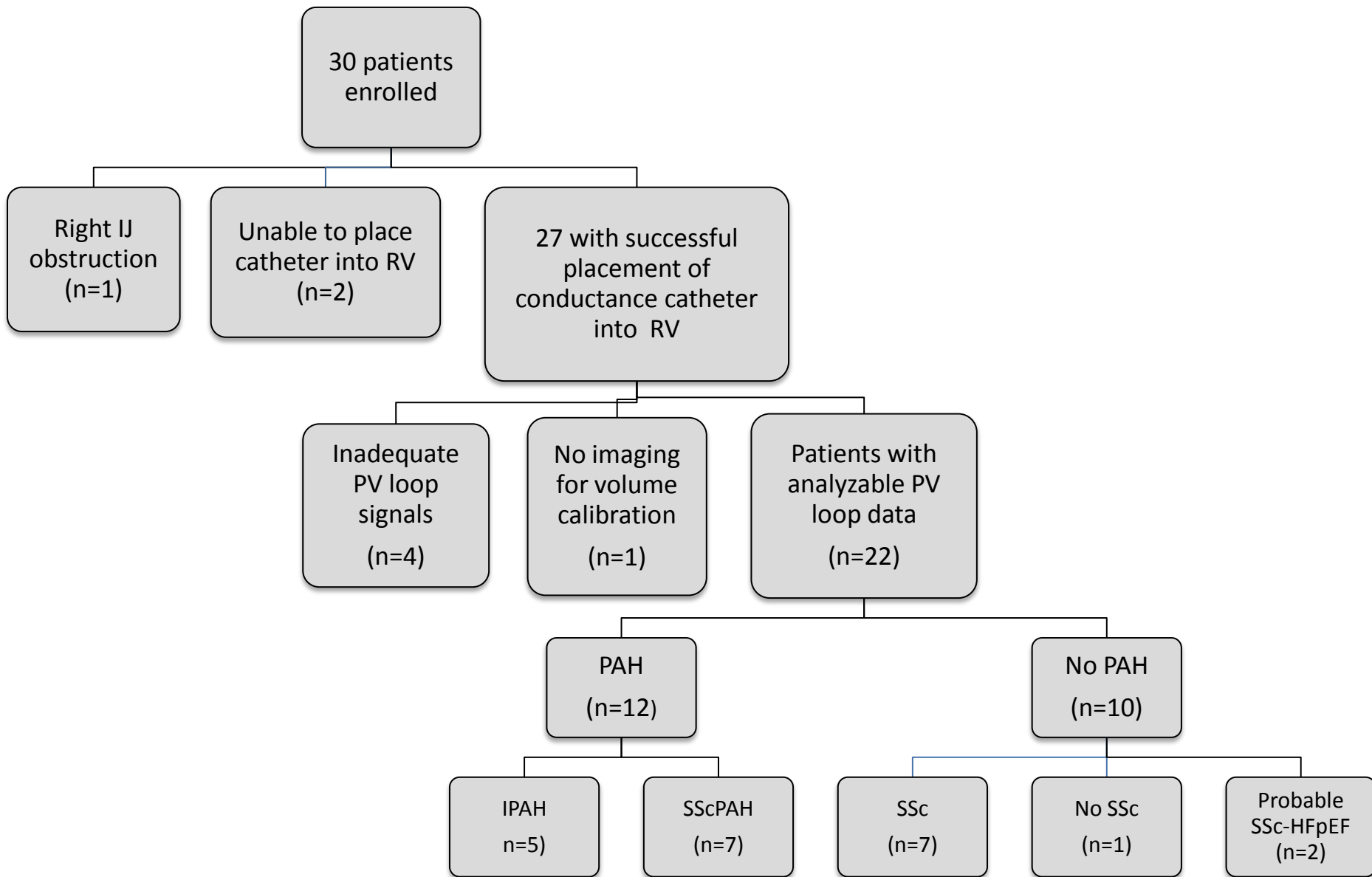
* Continuous variables shown as mean ± SD

Comparisons by student t-test or Mann-Whitney Rank Sum Test as appropriate

Online Supplement 2. Chronic Medications

Medication	IPAH (n=5)	SScPAH (n=7)	P-Value (IPAH vs. SScPAH)	SSc-no-PH (n=7)	P-Value (SScPAH vs. SSc-no-PAH)
PDE5A inhibitor	2 (40)	1 (14)	0.52	1 (14)	1.0
Endothelin Antagonist	2 (40)	0 (0)	0.15	0 (0)	1.0
Intravenous Prostacyclin	1 (20)	0 (0)	0.42	0 (0)	1.0
Inhaled or SQ Prostacyclin	2 (40)	0 (0)	0.15	0 (0)	1.0
Calcium Channel Blocker	1(20)	2 (29)	1.0	3 (43)	1.0
Loop Diuretic	3 (60)	1 (14)	0.22	2 (29)	1.0
Aldosterone Antagonist	3 (60)	1 (14)	0.22	2 (29)	1.0

PAH = Pulmonary Arterial Hypertension; PDE5A = Phosphodiesterase 5A; SQ = Subcutaneous
Comparisons by Fisher Exact Test



Online Supplement 3. Flow chart depicting patient enrollment in pressure-volume loop analysis. HFpEF = Heart failure with preserved ejection fraction.

Online Supplement 4. Change in Heart Rate with Valsalva maneuver. 2-4 beats are averaged just prior to initiation of Valsalva (baseline), during onset of initiation (phase 1-2), and after release maneuver (phase 3).