**Supplemental Methods**

**Diagnostic criteria for connective tissue diseases**

Each patient with systemic sclerosis (SSc), systemic lupus erythematosus (SLE), Sjögren's Syndrome (SS) and mixed connective tissue diseases (MCTD) fulfilled the 2013 American college of rheumatology (ACR) / European league against rheumatism (EULAR) collaborative SSc criteria [1], 1997 ACR revised criteria [2] or 2012 Systemic Lupus International Collaborating Clinics Classification criteria for SLE [3], the 2012 ACR classification criteria for SS [4], and at least one of the three MCTD criteria (Sharp, Alarcón-Segovia, or Kasukawa) [5], respectively. SSc were further classified into limited and diffuse forms of the disease [6].

**Right heart catheterization and clinical evaluation**

Pulmonary arterial (PA) capacitance was evaluated as follows: PA compliance = Stroke volume (SV) / PA pulse pressure. To calculate right ventricular end-diastolic (RVED) compliance, RVED pressure (RVEDP) and diastolic RV pressure (RVPd) were obtained, and the equation is as following: RVED compliance = SV / (RVEDP - RVPd).

**Echocardiography and pulmonary function test**

To evaluate tricuspid annular plane systolic excursion (TAPSE) in transthoracic echocardiography, the total displacement of the tricuspid annulus from end-diastole to end-systole was measured under the orientation of the M-mode cursor to the junction of the tricuspid valve plane with the RV free wall, according to the guidelines from the American Society of Echocardiography [7, 8].

Spirometry and the measurements of the diffusing capacity of the lung for carbon monoxide based on the single-breath method were performed (FUDAC-77(r) spirometer, Fukuda Denshi, Tokyo, Japan). The measurements of spirometry included forced vital capacity. The procedures and results of PFT according to the pulmonary function test guidelines of the Japanese Respiratory Society Guidelines [9], similar to those of the American Thoracic Society [10].

**Statistical analysis**

In univariate and multivariate Cox proportional logistic regression analysis, categorical variables including even no patients were excluded for the problem of separation. For univariate analysis, the factors of laboratory test, RHC and CMR analysis, PA stiffness metrics and PA-RV coupling metrics significantly associated with mortality due to PH from comparison between PH survivors and non-survivors were used. Associated demographics parameters including age, sex, PH classification (PAH vs. non-PAH), underlying disorder (SSc vs. non-SSc), ILD concomitance, were entered separately into the model. We used propensity score adjustment for each of the above risk factors, as previously described [11]. Adjustment by propensity score kept statistical power by the reduction of covariates into a single variable. When the adjusted effect of a risk factor was evaluated, the propensity score was computed through a binary logistic regression providing the predictability of having the risk factor as a function of the other risk factors. For continuous variables, the proportional odds logistic regression model was used to derive the propensity score.

**Supplemental Table 1.** Definition of right ventriculo-pulmonary arterial coupling measures and stiffness metrics

|  |  |  |
| --- | --- | --- |
| **Metrics** | **Measurement Description** | **Equation** |
| Ea, mmHg/ml/m2 | PA elastance | (mPAP - PAWP) / SVI |
| Ees, mmHg/ml/m2 | RV elastance | mPAP / RVESVI |
|  |  |  |
| Ees/Ea, ratio | PA-RV coupling metric | (mPAP / RVESVI) / [(mPAP - PAWP) / SVI] |
| CMR Ees/Ea, ratio | Non-invasive PA-RV coupling metric | SV / RVESV |
|  |  |  |
| PA capacitance, mL/mmHg | PA stiffness metric | SV / PA pulse pressure |
| RVED compliance, mL/mmHg | RV stiffness metric | SV / (RVEDP - RVPd) |
|  |  |  |
| PA relative area change, % | Noninvasive PA stiffness metric | (Maximal PA area - minimal PA area) / minimal PA area |
| PA distensibility, %/mmHg | PA stiffness metric | PA relative area change / PA pulse pressure |
| PA compliance, mm2/mmHg | PA stiffness metric | (maximal PA area - minimal PA area) / PA pulse pressure |
| PA elastic modulus, mmHg | PA stiffness metric | PA pulse pressure / (PA relative area change) |
| PA stiffness index β | PA stiffness metric | *Ln*{(sPAP / dPAP)} / (PA relative area change) |

Abbreviations: Ea, pulmonary arterial elastance; Ees, right ventricular elastance; PA, pulmonary artery; RV, right ventricle; mPAP, mean pulmonary arterial pressure; PAWP, pulmonary arterial wedge pressure; SVI, stroke volume index; RVESVI, right ventricular end-systolic volume index; CMR, cardiac magnetic resonance; RVED, right ventricular end-diastole; SV, stroke volume; RVEDP, right ventricular end-diastolic pressure; RVPd, diastolic right ventricular pressure; sPAP/dPAP, systolic/diastolic pulmonary arterial pressure

**References**

1. van den Hoogen F, Khanna D, Fransen J, Johnson SR, Baron M, Tyndall A, et al. 2013 classification criteria for systemic sclerosis: an American college of rheumatology/European league against rheumatism collaborative initiative. Ann Rheum Dis 2013;72:1747-55.

2. Hochberg MC. Updating the American college of rheumatology revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum 1997;40:1725-.

3. Petri M, Orbai A-M, Alarcón GS, Gordon C, Merrill JT, Fortin PR, et al. Derivation and validation of the Systemic Lupus International Collaborating Clinics classification criteria for systemic lupus erythematosus. Arthritis Rheum 2012;64:2677-86.

4. Shiboski SC, Shiboski CH, Criswell L, Baer A, Challacombe S, Lanfranchi H, et al. American College of Rheumatology classification criteria for Sjogren's syndrome: a data-driven, expert consensus approach in the Sjogren's International Collaborative Clinical Alliance cohort. Arthritis Care Res (Hoboken) 2012;64:475-87.

5. Ortega-Hernandez OD, Shoenfeld Y. Mixed connective tissue disease: an overview of clinical manifestations, diagnosis and treatment. Best Pract Res Clin Rheumatol 2012;26:61-72.

6. LeRoy EC, Medsger TA, Jr. Criteria for the classification of early systemic sclerosis. J Rheumatol 2001;28:1573-6.

7. Rudski LG, Lai WW, Afilalo J, Hua L, Handschumacher MD, Chandrasekaran K, et al. Guidelines for the echocardiographic assessment of the right heart in adults: a report from the American Society of Echocardiography endorsed by the European Association of Echocardiography, a registered branch of the European Society of Cardiology, and the Canadian Society of Echocardiography. J Am Soc Echocardiogr 2010;23:685-713; quiz 86-8.

8. Bolca O, Gungor B, Gurkan U, Yilmaz H. Assessment of right ventricular systolic function in patients with pulmonary hypertension. J Am Soc Echocardiogr 2012;25:804.

9. [Guideline of respiratory function tests--spirometry, flow-volume curve, diffusion capacity of the lung]. Nihon Kokyuki Gakkai Zasshi 2004;Suppl:1-56.

10. Culver BH, Graham BL, Coates AL, Wanger J, Berry CE, Clarke PK, et al. Recommendations for a Standardized Pulmonary Function Report. An Official American Thoracic Society Technical Statement. Am J Respir Crit Care Med 2017;196:1463-72.

11. Patterson JA, Stuart EA, Ford JB. Use of propensity score methods to address adverse events associated with the storage time of blood in an obstetric population: a comparison of methods. BMC Res Notes 2016;9:367.