**Associated pulmonary hypertension is an independent contributor to exercise intolerance in chronic fibrosing interstitial pneumonias**

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**SUPPLEMENTAL MATERIAL**

**e-Table 1**.Relationship between 6MWD (m), demographic, functional, hemodynamic and radiographic variables in all patients (n=72).

|  |  |  |
| --- | --- | --- |
|  | **Correlation Coefficient (r/rho)** | **pvalue** |
| **Demographics** | | |
| **Age** | -0.53 | <0.0001 |
| **BMI** | -0.30 | 0.0135 |
| **Hemodynamics** | | |
| **mPAP** | -0.32 | 0.0066 |
| **CO** | 0.17 | n.s. |
| **PVR** | -0.28 | 0.0253 |
| **Pulmonary Function** | | |
| **FVC** (% pred) | -0.27 | 0.0253 |
| **DLCO** (% pred) | -0.02 | n.s. |
| **Radiographic Data** | | |
| **PA Size** | -0.15 | n.s. |
| **PA:A Ratio** | -0.22 | n.s. |
| **HRCT Fibrosis** | 0.11 | n.s. |

Abbreviations: A: ascending aorta, BMI: body mass index, CO: cardiac output, DLCO: carbon monoxide diffusion capacity, FVC: forced vital capacity, HRCT: high-resolution computed tomography, mPAP: mean pulmonary arterial pressure, PA: pulmonary artery, PVR: pulmonary vascular resistance, 6MWD: six-minute walk distance.

**e-Table 2**. Relationship between 6MWD (% pred), demographic, functional, hemodynamic and radiographic variables in patients with APH only (n=55).

|  |  |  |
| --- | --- | --- |
|  | **Correlation Coefficient (r/rho)** | **pvalue** |
| **Pulmonary function** | | |
| **FVC** (% pred) | -0.14 | n.s. |
| **DLCO** (% pred) | -0.03 | n.s. |
| **Hemodynamics** | | |
| **mPAP** | -0.31 | 0.0221 |
| **CO** | 0.21 | n.s. |
| **PVR** | -0.33 | 0.0199 |
| **CT measurements** | | |
| **PA Size** | -0.13 | n.s. |
| **PA:A Ratio** | -0.03 | n.s. |
| **HRCT Fibrosis** | -0.01 | n.s. |

Abbreviations: A: ascending aorta, APH: associated pulmonary hypertension, BMI: body mass index, CO: cardiac output, DLCO: carbon monoxide diffusion capacity, FVC: forced vital capacity, HRCT: high-resolution computed tomography, mPAP: mean pulmonary arterial pressure, PA: pulmonary artery, PVR: pulmonary vascular resistance, 6MWD: six-minute walk distance.

**e-Table 3**. Independent, significant predictors of 6MWD (% pred) selected by stepwise multiple regression analysis. Patients with APH only (n=55). HRCT fibrosis score, FVC, DLCO, left heart dysfunction, coronary artery disease, sleep apnea, pulmonary rehabilitation, presence of obstructive defect and concomitant emphysema were not significant.

|  |  |  |
| --- | --- | --- |
| **Covariables** | **F ratio** | **p value** |
| **PVR** | 4.52 | 0.0387 |

Abbreviations: APH: associated pulmonary hypertension, DLCO: carbon monoxide diffusion capacity, FVC: forced vital capacity, HRCT: high-resolution computed tomography, PVR: pulmonary vascular resistance, 6MWD: six-minute walk distance.

**e-Table 4.** Independent, significant predictors of 6MWD (m) selected by stepwise multiple regression analysis. All patients (n=72). HRCT fibrosis score, FVC, DLCO, left heart dysfunction, coronary artery disease, sleep apnea, pulmonary rehabilitation, presence of obstructive defect and concomitant emphysema were not significant.

|  |  |  |  |
| --- | --- | --- | --- |
| **Covariables** | **F ratio** | **p-value** | **VIF vs PVR** |
| **Age** | 18.70 | 0.0001 | 1.03 |
| **PVR** | 4.69 | 0.0343 |  |
| **BMI** | 3.91 | 0.0524 | 1.03 |

Abbreviations: BMI: body mass index, DLCO: carbon monoxide diffusion capacity, FVC: forced vital capacity, HRCT: high-resolution computed tomography, PVR: pulmonary vascular resistance, VIF: variance inflation factor, 6MWD: six-minute walk distance.

**e-Table 5A**. Independent, significant predictors of 6MWD (% pred) selected by stepwise multiple regression analysis. Patients with normal wedge pressure (≤15 mmHg) only (n=62). HRCT fibrosis score, FVC, DLCO, left heart dysfunction, coronary artery disease, sleep apnea, pulmonary rehabilitation, presence of obstructive defect and concomitant emphysema were not significant.

|  |  |  |
| --- | --- | --- |
| **Covariables** | **F ratio** | **p value** |
| **PVR** | 4.26 | 0.0440 |

Abbreviations: DLCO: carbon monoxide diffusion capacity, FVC: forced vital capacity, HRCT: high-resolution computed tomography, PVR: pulmonary vascular resistance, 6MWD: six-minute walk distance.

**e-Table 5B**. Independent, significant predictors of 6MWD (m) selected by stepwise multiple regression analysis. Patients with normal wedge pressure (≤15 mmHg) only (n=62). HRCT fibrosis score, FVC, DLCO, left heart dysfunction, coronary artery disease, sleep apnea, pulmonary rehabilitation, presence of obstructive defect and concomitant emphysema were not significant.

|  |  |  |
| --- | --- | --- |
| **Covariables** | **F ratio** | **p value** |
| **Age** | 16.90 | 0.0001 |
| **BMI** | 4.69 | 0.0349 |
| **PVR** | 4.38 | 0.0412 |

Abbreviations: BMI: body mass index, DLCO: carbon monoxide diffusion capacity, FVC: forced vital capacity, HRCT: high-resolution computed tomography, PVR: pulmonary vascular resistance, 6MWD: six-minute walk distance.

**e-Table 6**. Independent, significant predictors of MRC dyspnea score (MRCDS) selected by stepwise multiple regression analysis. Patients with APH only (n=55). HRCT fibrosis score, FVC, DLCO, left heart dysfunction, coronary artery disease, sleep apnea, pulmonary rehabilitation, presence of obstructive defect and concomitant emphysema were not significant.

|  |  |  |
| --- | --- | --- |
| **Covariables** | **F ratio** | **p value** |
| **mPAP** | 5.55 | 0.0226 |

Abbreviations: APH: associated pulmonary hypertension, DLCO: carbon monoxide diffusion capacity, FVC: forced vital capacity, HRCT: high-resolution computed tomography, mPAP: mean pulmonary arterial pressure, MRCDS: Medical Research Council dyspnea score.