Disease Activity Is Related to Acute Response to Vasodilator in Pulmonary Artery Hypertension Associated With Systemic Lupus Erythematosus

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**Background:** No previous study has been done on whether systemic lupus erythematosus (SLE) disease activity is related to the hemodynamics and right ventricular (RV) function in patients with SLE-associated pulmonary artery hypertension (SLE-APAH).

**Methods and Results:** This study prospectively recruited 54 patients (mean age, 32.8 ± 8.4 years; 92.6% female) with SLE-APAH, including 34 patients with SLE disease activity index (SLEDAI) <5 (low score) and 20 with SLEDAI ≥5 (high score). All patients underwent right heart catheterization and iloprost inhalation, and echocardiography was performed before and immediately after iloprost inhalation. There was no difference in baseline mean pulmonary artery pressure (mPAP) between the 2 groups; pulmonary vascular resistance (PVR) was significantly higher and cardiac index was significantly lower in the low-SLEDAI group. The patients with low SLEDAI had larger RV size and worse RV systolic function on echocardiography. After iloprost inhalation, the patients with low SLEDAI had a greater decrease in mPAP and PVR than those with high SLEDAI, while significantly increased RV systolic function was found only in the low-SLEDAI group.

**Conclusions:** SLE activity is related to hemodynamics and RV function in SLE-APAH patients, and those with low SLEDAI might have better acute response to vasodilator inhalation. *(Circ J 2014; 78: 1240–1244)*

**Key Words:** Disease activity; Hemodynamics; Pulmonary artery hypertension; Right heart function; Systemic lupus erythematosus

Pulmonary artery hypertension (PAH) is a potentially life-threatening complication of systemic lupus erythematosus (SLE). The onset of PAH associated with SLE (SLE-APAH) does not correlate with the disease duration, and patients may even present with PAH before SLE diagnosis. Vasodilators such as inhaled iloprost have been suggested in the treatment of SLE-APAH, but no study has investigated whether the hemodynamic parameters and acute hemodynamic response to inhaled iloprost, as determined on right heart catheterization (RHC), are related to SLE activity in SLE-APAH patients.

Right heart dysfunction is very important in the progress of PAH, and right heart function assessed on echocardiography is also an important determinant of prognosis in PAH patients. Advanced echocardiographic techniques could provide precise quantitative information on complex right ventricular (RV) wall motion and alternative indicators of hemodynamic changes in PAH patients. In a recent study, right heart function assessed on tissue Doppler imaging (TDI) was found to be related to the response to vasodilator in PAH associated with connective tissue disease (CTD), but little is known about whether right heart function, and its reaction to vasodilator, are related to SLE activity in SLE-APAH patients.

This study was designed to evaluate the hypothesis that SLE disease activity is related to the acute response (including the hemodynamic response and right heart function change) to in-
inhaled pulmonary vasodilator in SLE-APAH patients.

**Methods**

**Ethics Statement**

The study protocol was reviewed and approved by the Ethics Committee of Peking Union Medical College Hospital, Chinese Academy of Medical Sciences (Beijing, China), and all patients provided written informed consent.

**Patients**

Between February 2010 and February 2012, patients who underwent RHC for SLE-APAH were enrolled in the study. PAH was defined as mean pulmonary arterial pressure (mPAP) >25 mmHg with a pulmonary capillary wedge pressure (PCWP) ≤15 mmHg and pulmonary vascular resistance (PVR) >3 Wood units on RHC. SLE was diagnosed according to the 1982 revised American Rheumatism Association (ARA) criteria. Patients with previous atrial fibrillation, coronary artery disease, valvular disease and impaired left ventricular (LV) systolic function (defined as LV ejection fraction [LVEF] <50%) were excluded. SLE activity was assessed using the SLE disease activity index (SLEDAI), and the patients with SLEDAI <5 were considered as having low SLE activity.

**Hemodynamic Measurements and Inhaled Vasodilator**

RHC hemodynamic assessments were obtained before and after iloprost inhalation. An 8.5-F introducer sheath was placed in the right internal jugular vein or the right subclavian vein, and a 6-lumen 8-F Swan-Ganz catheter (Edwards Lifesciences World Trade, Irvine, CA USA) was advanced into the pulmonary artery. Mean systemic blood pressure (SBPmean), mean right atrial pressure (mRAP), mPAP, and PCWP were measured at baseline and after vasodilator drug treatment. Cardiac output (CO) was measured in triplicate with the thermodilution technique (Edwards Lifesciences World Trade). The cardiac index (CI) was calculated using CO indexed to body surface area. PVR was calculated using the standard hemodynamic formulas as follows: PVR=(mPAP–PCWP)/CO.

After the baseline measurement of hemodynamic parameters, 20 µg iloprost (Ventavis1; Bayer-Schering Pharma, Berlin, Germany) was delivered with a PARI LC STAR nebulizer (PARI, Starnberg, Germany) driven by a PARI TurboBOY-N compressor (PARI) for 10–15 min. Another complete set of hemodynamic parameters was obtained at the end of inhalation.

**Echocardiography**

All subjects underwent echocardiography, including M-mode, 2-D conventional Doppler and TDI echocardiography, before and immediately after iloprost inhalation using commercially available equipment (Vivid I; GE Vingmed Ultrasound). All examinations were performed and reviewed by cardiologists with advanced training in echocardiography.

RA area (RAA), inferior vena cava (IVC), and tricuspid anular plane systolic excursion (TAPSE) were measured according to the American Society of Echocardiography’s Guidelines. RV end-diastolic area (RVEDA), RV end-systolic area (RVESA) and RV fractional area change (RVFAC) were assessed in the apical 4-chamber view. LV end-diastolic volume (LVEDV), LV end-systolic volume (LVESV) and LVEF were assessed using the modified biplane Simpson’s equation in the apical 4- and 2-chamber views. TDI was performed in the apical 4-chamber view to assess the long-axis motion of the heart. Pulse TDI sample volume was placed at the lateral tricuspid valve (TV) annulus to obtain the peak systolic velocity of the lateral TV annulus (TVS') and early diastolic velocity of the lateral TV annulus (TVE') and later diastolic velocity of the lateral TV annulus (TVA'; Figure). The TV closure–opening time (TCO) was measured on pulsed TDI, which encompassed isovolumic contraction time, ejection time (ET), and isovolumic relaxation time. The RV performance index (RVMPI) was calculated according the following formula: RVMPI=(TCO–ET)/ET.

Tricuspid inflow was assessed on pulsed-wave Doppler echocardiography in the apical 4-chamber view. The Doppler beam was aligned parallel to the direction of flow and a 1–2 mm sample volume was placed between the tips of the TV during diastole. From the inflow profile, the E- and A-wave velocity of the TV was obtained. The ratio of E/A from TV was calculated.

During echocardiography, at least 3 consecutive beats were stored and the images were digitized and analyzed off-line using...
patients with SLEDAI ≥5 (high score). The patients were divided into low-SLEDAI and high-SLEDAI groups. Baseline clinical characteristics, hemodynamic parameters and echocardiographic data are listed in Table 1.

There was no difference in age, gender, baseline SBP mean or heart rate between the 2 groups. No difference could be found in the prevalence of accompanying CTD, Raynaud phenomenon, positive anti-U1 ribonucleoprotein (RNP) antibody and pleural effusions, or the duration of PAH between the 2 groups. The patients with low SLEDAI, however, tended to have longer 6-min walk distance (P=0.05). As for the baseline hemodynamic parameters, although there was no difference in mPAP and PCWP between the 2 groups, PVR and mRAP were significantly higher and CI was significantly lower in the patients with low SLEDAI.

With regard to the baseline echocardiographic data, larger RVEDA, smaller LVEDV and IVC were found in the patients with low SLEDAI. No difference could be found in LVEF and RAA between the 2 groups. The low-SLEDAI group had lower TVS’ and TAPSE, reflecting RV longitudinal systolic function.

Table 1. Baseline Patient Characteristics vs. SLEDAI

<table>
<thead>
<tr>
<th>SLEDAI &lt;5 (n=34)</th>
<th>SLEDAI ≥5 (n=20)</th>
<th>P-value</th>
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<tbody>
<tr>
<td>Age (years)</td>
<td>33.8±7.1</td>
<td>31±10.2</td>
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<tr>
<td>Gender (female)</td>
<td>32 (94.1)</td>
<td>18 (90)</td>
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<tr>
<td>Accompanying CTD</td>
<td>8 (23.5)</td>
<td>6 (30)</td>
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<tr>
<td>Raynaud phenomenon</td>
<td>14 (41.2)</td>
<td>8 (40)</td>
</tr>
<tr>
<td>Positive anti-U1 RNP antibody</td>
<td>20 (58.8)</td>
<td>12 (60)</td>
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<tr>
<td>Pleural effusions</td>
<td>22 (64.7)</td>
<td>14 (70)</td>
</tr>
<tr>
<td>6MWD (m)</td>
<td>442.6±83</td>
<td>403±40.1</td>
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<tr>
<td>Duration of PAH (months)</td>
<td>7.3±13.1</td>
<td>6.0±5.9</td>
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<td>HR (beats/min)</td>
<td>80.9±12.1</td>
<td>77.4±13.7</td>
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<td>SBP mean (mmHg)</td>
<td>86.6±7.4</td>
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<th>Hemodynamic parameters</th>
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<td>mPAP (mmHg)</td>
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<td>PCWP (mmHg)</td>
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<tr>
<td>CO (L/min)</td>
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<tr>
<td>PVR (Wood units)</td>
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<tr>
<td>CI (L · min⁻¹ · m⁻²)</td>
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<tr>
<th>Echocardiographic data</th>
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<tr>
<td>LVEDV (ml)</td>
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<tr>
<td>LVEF (biplane, %)</td>
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<tr>
<td>RAA (cm²)</td>
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<tr>
<td>RVEDA (cm²)</td>
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<tr>
<td>TVS’ (cm/s)</td>
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<tr>
<td>TAPSE (mm)</td>
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<tr>
<td>RVFAC (%)</td>
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<tr>
<td>RVMPI (%)</td>
</tr>
<tr>
<td>TVE (cm/s)</td>
</tr>
<tr>
<td>TVA (cm/s)</td>
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<tr>
<td>TV-E/A</td>
</tr>
<tr>
<td>TVE’ (cm/s)</td>
</tr>
<tr>
<td>IVC (mm)</td>
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</tbody>
</table>

Data given as mean±SD or n (%).

6MWD, 6-min walk distance; CI, cardiac index; CO, cardiac output; CTD, connective tissue disease; HR, heart rate; IVC, inferior vena cava; LVEDV, left ventricle end-diastolic volume; LVEF, left ventricular ejection fraction; mPAP, mean pulmonary arterial pressure; mRAP, mean right atrial pressure; PCWP, pulmonary capillary wedge pressure; PVR, pulmonary vascular resistance; RAA, right atrial area; RNP, ribonucleoprotein; RVEDA, right ventricular end-diastolic area; RVFAC, right ventricle fractional area change; RVMPI, right ventricle performance index; SBP mean, mean systemic blood pressure; SLEDAI, systemic lupus erythematosus disease activity index; TAPSE, tricuspid annular plane systolic excursion; TVA, tricuspid valve peak A-wave velocity; TVE, tricuspid valve peak E-wave velocity; TVE’, early diastolic velocity of lateral tricuspid valve annulus; TV-E/A, ratio of tricuspid valve peak E-wave to peak A-wave velocity; TVS’, peak systolic velocity of lateral tricuspid valve annulus.

Statistical Analysis

The difference in means between the 2 groups was examined using independent or paired t-test and categorical variables were analyzed using chi-squared test or Fisher’s exact test. SPSS version 13.0 (SPSS, Chicago, IL, USA) was used for calculations. All data are expressed as mean±SD. P<0.05 was considered statistically significant.

Results

A total of 54 patients (mean age, 32.8±8.4 years; 92.6% female) were enrolled in the study. Fourteen patients had other CTD, including 8 patients with systemic sclerosis, 5 patients with Sjogren’s syndrome and 1 patient with rheumatoid arthritis. There were 34 patients with SLEDAI <5 (low score) and 20 patients with SLEDAI ≥5 (high score). The patients were divided into low-SLEDAI and high-SLEDAI groups. Baseline clinical characteristics, hemodynamic parameters and echocardiographic data are listed in Table 1.

There was no difference in age, gender, baseline SBP mean or heart rate between the 2 groups. No difference could be found in the prevalence of accompanying CTD, Raynaud phenomenon, positive anti-U1 ribonucleoprotein (RNP) antibody and pleural effusions, or the duration of PAH between the 2 groups. The patients with low SLEDAI, however, tended to have longer 6-min walk distance (P=0.05). As for the baseline hemodynamic parameters, although there was no difference in mPAP and PCWP between the 2 groups, PVR and mRAP were significantly higher and CI was significantly lower in the patients with low SLEDAI.

With regard to the baseline echocardiographic data, larger RVEDA, smaller LVEDV and IVC were found in the patients with low SLEDAI. No difference could be found in LVEF and RAA between the 2 groups. The low-SLEDAI group had lower TVS’ and TAPSE, reflecting RV longitudinal systolic function.
and tended to have lower TVE’ (P=0.05), reflecting RV diastolic function. The 2 groups had similar RVFAC, RVMPI, TV-E/A and TVE.

The hemodynamic and echocardiographic data before and immediately after iloprost inhalation are given in Table 2. After the use of vasodilator, SBPmean, mPAP and PVR decreased significantly and CI increased significantly in both groups. The RV systolic parameters assessed on echocardiography (TVS’, TAPSE and RVFAC) indicated a different reaction to the vasodilator by the 2 groups; TVS’ and TAPSE increased significantly in the low-SLEDAI group but no such change was seen in the high-SLEDAI group. RVFAC remained unchanged in both groups.

We also compared the change in hemodynamic parameters and RV systolic function after iloprost inhalation between the 2 groups and the results are listed in Table 3. Although mPAP and PVR decreased significantly after the use of vasodilator in both groups (Table 2), the patients with low SLEDAI had a greater decrease in mPAP and PVR after iloprost inhalation than did those with high SLEDAI. Iloprost inhalation increased CO and CI with the same amplitude in both groups. The improvement of TVS and TAPSE, which reflected RV longitudinal systolic function, was more obvious in the low-SLEDAI group.

Discussion

To our knowledge, this is the first study to confirm that SLE disease activity is related to the characteristics of SLE-APAH. The main findings are as follows: even at a similar level of PAH, (1) low-SLEDAI SLE-APAH patients had higher PVR, lower CO and worse RV longitudinal systolic function than high-SLEDAI patients; and (2) low-SLEDAI patients might benefit more from prostanoid inhalation, including more significantly decreased mPAP and more obviously improved RV longitudinal systolic function.

The causal relationship between SLE and PAH has not been established, but the various elements of SLE, such as vasoconstriction, pulmonary vasculitis, and interstitial lung disease, can lead to endothelial and smooth muscle proliferation and damage of the pulmonary vasculature, resulting in PAH. Besides mechanisms similar to those in idiopathic PAH, mechanisms involving inflammation and autoimmunity may play a significant role in the pathogenesis or progression of SLE-APAH. Some studies have suggested an imbalance between vasoconstrictors and vasodilators in SLE-APAH. According to the present results, there might be difference in the principal mechanism of SLE-APAH according to SLE activity. In the present study, at a similar level of mPAP, lower SLEDAI was related to higher PVR, larger RV size, and worse RV longitudinal systolic function, which might indicate severer PAH in these patients. But in these low-SLEDAI patients we could also observe better response to the vasodilator, meaning that reversible vasoconstriction might make a greater contribution to the pathogenesis of PAH in these patients. Given that high SLEDAI suggests active lupus, it is reasonable to hypothesize that immune and inflammatory injury might be an important mechanism in the pathogenesis of SLE-APAH in these patients. The immune and inflammatory injury, but not reversible vasoconstriction, might explain the relatively lower response to the vasodilator in these patients. In fact, in a recent study, Sanchez et al reported that 8 of 28 patients with CTD-associated PAH, including 5 of 13 patients with SLE-APAH, responded clinically and hemodynamically after immunosuppressive therapy without combination therapy targeting PAH.

Table 2. Response to Iloprost Inhalation vs. SLEDAI

<table>
<thead>
<tr>
<th></th>
<th>SLEDAI &lt;5 (n=34)</th>
<th>SLEDAI ≥5 (n=20)</th>
<th>P-value</th>
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<tbody>
<tr>
<td>HR (beats/min)</td>
<td>Before</td>
<td>After</td>
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<td></td>
<td>80.9±12.1</td>
<td>78.8±11.6</td>
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<td></td>
<td>77.4±13.7</td>
<td>78.1±10.5</td>
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<tr>
<td>SBPmean (mmHg)</td>
<td>Before</td>
<td>After</td>
<td></td>
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<tr>
<td></td>
<td>86.6±7.4</td>
<td>79.1±9.1</td>
<td>&lt;0.01</td>
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<td></td>
<td>83.8±11.2</td>
<td>77.5±10.5</td>
<td>&lt;0.01</td>
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<tr>
<td>mPAP (mmHg)</td>
<td>Before</td>
<td>After</td>
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<tr>
<td></td>
<td>46.8±14.3</td>
<td>40.4±15.2</td>
<td>&lt;0.01</td>
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<td></td>
<td>43.3±10.4</td>
<td>38.0±10.9</td>
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<tr>
<td>CO (L/min)</td>
<td>Before</td>
<td>After</td>
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<tr>
<td></td>
<td>4.0±1.2</td>
<td>4.3±1.2</td>
<td>&lt;0.01</td>
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<td>4.8±1.3</td>
<td>5.1±1.3</td>
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<td>CI (L·min⁻¹·m⁻²)</td>
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<td>After</td>
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<td></td>
<td>2.3±0.8</td>
<td>2.5±0.8</td>
<td>&lt;0.01</td>
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<tr>
<td></td>
<td>3.0±0.8</td>
<td>3.2±0.8</td>
<td>&lt;0.01</td>
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<td>PVR (Wood units)</td>
<td>Before</td>
<td>After</td>
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<tr>
<td></td>
<td>12.4±8.2</td>
<td>9.4±6.8</td>
<td>&lt;0.01</td>
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<tr>
<td></td>
<td>8.7±3.5</td>
<td>7.0±2.8</td>
<td>&lt;0.01</td>
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<tr>
<td>TVS’ (cm/s)</td>
<td>Before</td>
<td>After</td>
<td></td>
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<tr>
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<td>10.4±2.4</td>
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<td>12.2±2.0</td>
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<td>TAPSE (mm)</td>
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<td>17.6±2.8</td>
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<td>RVFAC (%)</td>
<td>Before</td>
<td>After</td>
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<tr>
<td></td>
<td>0.31±0.11</td>
<td>0.31±0.10</td>
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<tr>
<td></td>
<td>0.36±0.14</td>
<td>0.36±0.08</td>
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Data given as mean±SD. Abbreviations as in Table 1.

Table 3. Changes After Iloprost Inhalation

<table>
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<tr>
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<th>SLEDAI &lt;5 (n=34)</th>
<th>SLEDAI ≥5 (n=20)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Decreased mPAP (mmHg)</td>
<td>Before</td>
<td>After</td>
<td></td>
</tr>
<tr>
<td></td>
<td>6.4±3.0</td>
<td>4.4±2.7</td>
<td>0.02</td>
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<tr>
<td></td>
<td>0.3±0.04</td>
<td>0.3±0.05</td>
<td>0.94</td>
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<tr>
<td>Increased CO (L/min)</td>
<td>Before</td>
<td>After</td>
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<td></td>
<td>0.2±0.2</td>
<td>0.2±0.3</td>
<td>0.7</td>
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<tr>
<td></td>
<td>1.6±1.7</td>
<td>&lt;0.01</td>
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<tr>
<td>Decreased PVR (Wood units)</td>
<td>Before</td>
<td>After</td>
<td></td>
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<tr>
<td></td>
<td>1.5±2.1</td>
<td>-0.2±1.8</td>
<td>&lt;0.01</td>
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<tr>
<td></td>
<td>0.2±1.9</td>
<td>&lt;0.01</td>
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<tr>
<td>Improved TVS’ (cm/s)</td>
<td>Before</td>
<td>After</td>
<td></td>
</tr>
<tr>
<td></td>
<td>2.6±3.4</td>
<td>-0.2±1.9</td>
<td>&lt;0.01</td>
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<tr>
<td></td>
<td>0±14.0</td>
<td>&lt;0.01</td>
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<tr>
<td>Improved RVFAC (%)</td>
<td>Before</td>
<td>After</td>
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<tr>
<td></td>
<td>1.0±7.0</td>
<td>0±14.0</td>
<td>0.92</td>
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Data given as mean±SD. Abbreviations as in Table 1.
The authors defined response to immunosuppressive therapy as maintenance of New York Heart Association functional class I or II with sustained hemodynamic improvement after at least 1 year of immunosuppressive therapy, but the authors did not present the SLEDAI score. That study and the present results suggest that the inflammatory mechanisms might contribute to the genesis of SLE-APAH, which could partly explain the poor response to vasodilator in the present study and the good response to immunosuppressive therapy in the Sanchez et al study in some patients with SLE-APAH.

The present results show that patients with high SLEDAI had less severe SLE than patients with low SLEDAI in terms of CIC and PVR. This suggests that these patients could have been in an earlier phase of the disease, but the mean delay between PAH diagnosis and RHC was identical in both groups.

In the present study, the different RV systolic parameters responded differently to the use of vasodilator. As is known, the RV has superficial circumferential muscle fibers responsible for its inward bellows movement, as well as inner longitudinal fibers that result in the base-to-apex contraction. Compared with the LV, the base-to-apex shortening plays a greater role in RV emptying. Our previous study showed that TAPSE and TVS', which reflect the velocity and distance of the TV annulus base-to-apex movement, were more sensitive to impairment of right heart function in PAH patients; the present study has further demonstrated that TAPSE and TVS' are more sensitive in both reflecting differing RV function impairment in SLE-APAH patients according to SLEDAI, and reflecting the improvement of RV function after the use of vasodilator.

**Study Limitations**

There were several potential limitations to the present study. First, the subject group was relatively small. We focused on SLE-APAH patients, however, all of whom underwent RHC and advanced echocardiography before and immediately after vasodilator inhalation, and even with these numbers we were able to demonstrate the differing hemodynamic and RV function characteristics with different SLEDAI. Second, TDI measurements of the TV annulus are limited by the angle dependency of the technique, but with careful adjustment of the beam and gain settings to avoid aliasing, reliable measurement of tissue velocities of the TV annulus can be made. Third, the cross-sectional design of the study limited the ability to examine long-term reaction to the vasodilator in SLE-APAH patients according to SLEDAI.

**Conclusions**

SLE activity is related to the hemodynamic parameters and RV function in SLE-APAH patients, and those patients with low SLEDAI might have better acute response to vasodilator inhalation.

**References**


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