from: Actionel, Astra Zeneca, BMS, GSK, Janssen, UCB, Alberta Hoi Speakers bureau: UCB, Janssen, Sandzo, Eli Lilly, Consultant of: Abbvie, GSK, Grant/ research support from: AstraZeneca, GSK, BMS, Janssen, and Merck Serono, Eric F. Morand Speakers bureau: AstraZeneca, EMD Serono, Gilead, Consultant of: AstraZeneca, BristolMyersSquibb, Biogen, Eli Lilly, EMD Serono, Novartis, Grant/research support from: AbbVie, Amgen, AstraZeneca, Bristol-Myers, Squibb, Biogen, Eli Lilly, EMD Serono, Genentech, GSK, Janssen, UCB.

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Table 1. Baseline characteristics of SLE-PAH and IPAH patients

<table>
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<th>Trait</th>
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<th>p-value</th>
</tr>
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<tbody>
<tr>
<td>Age at enrollment, yrs</td>
<td>55.6±4.1</td>
<td>6.2±4.6</td>
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<tr>
<td>Female, No.(%)</td>
<td>2.2±0.9</td>
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<tr>
<td>NT-proBNP, ng/L</td>
<td>56.3±16.8</td>
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</tr>
<tr>
<td>MPA, mmHg</td>
<td>56.3±16.8</td>
<td>33.2±6.5</td>
</tr>
<tr>
<td>PVR, WU</td>
<td>14.9±3.9</td>
<td>13.7±3.5</td>
</tr>
<tr>
<td>TLC, (predicted)</td>
<td>60.1±11.8</td>
<td>60.1±11.3</td>
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| PULMONARY ARTERIAL HYPERTENSION: A 10-YEAR MULTICENTER COHORT STUDY

Keywords: Systemic lupus erythematosus, Cardiovascular disease, Treat to target

X. Dong1, J. Qian1, J. L. Zhao1, Q. Wang1, M. Li1, X. Zeng1, 1Peking Union Medical College Hospital, Department of Rheumatology and Clinical Immunology, Beijing, China

Background: Previous studies have described improved survival in systemic sclerosis (SSc) associated pulmonary arterial hypertension (PAH) [1-12], yet it is unclear whether survival of systemic lupus erythematosus (SLE) associated PAH has improved.

Objectives: To describe the CSTAR prospective cohort of SLE-PAH patients, explore changes in characteristics, treatment and 5-year survival in the last decade, and possible reasons for survival change.

Methods: A multicenter prospective cohort of SLE-PAH was established, and divided into cohort A (2011.6-2015.5) and cohort B (2016.6-2021.5) according to the date of their baseline right heart catheterization (RHC). Another singlecenter cohort of idiopathic pulmonary arterial hypertension (IPAH) was consecutively recruited as control to describe the baseline characteristic and survival of SLE-PAH patients simultaneously. Disease characteristics, treatment regimen, and all-cause mortality were compared between cohort A and cohort B. Multi-variable cox regression was used to analyze association between treatment goal achievement and survival.

Results: A total of 610 SLE-PAH and 104 IPAH patients were enrolled. Overall, SLE-PAH patients were younger, predominantly female, had lower NT-proBNP level, better function status, better hemodynamic, and higher 5-year survival than IPAH (81.2% vs 56.0%, p<0.001). Compared with cohort A, patients in cohort B showed lower mPAP and PVR, higher CI, and were more likely to receive intensive immunosuppressants and PAH-targeted medication. 5-year survival rate was also higher in cohort B (88.1% vs 72.9%, p=0.01). In multivariable Cox regression, treatment goal achievement of PAH (HR 0.31, 95% CI 0.12-0.81, p=0.017) and reaching lupus low disease activity state (LLDAS) (HR 0.23, 95% CI 0.08-0.67, p=0.007) were both independently associated with a lower mortality.

Conclusion: This is the largest multicenter prospective SLE-PAH cohort to describe disease characteristics and prognosis. This study showed that survival has improved significantly for SLE-PAH. Early detection of PAH in SLE patients, achieving PAH treatment goal as well as LLDAS for SLE contributed to survival improvement.

REFERENCES:

Acknowledgements: NIL.

Disclosure of Interests: None Declared.

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OP0228

COEXISTING TUBULOINTERSTITIAL INFLAMMATION AND DAMAGE IS A RISK FACTOR FOR CHRONIC KIDNEY DISEASE IN PATIENTS WITH LUPUS NEPHRITIS: RESULTS FROM THE KORNET REGISTRY

Keywords: Systemic lupus erythematosus

D. J. Park1, S. E. Choi1, J. H. Kang1, S. S. Lee1, 1Chonnam National University Medical School and Hospital, Rheumatology, Gwangju, Korea, Rep. of (South Korea)

Background: An increasing body of evidence suggests a prognostic role of tubulointerstitial lesions in patients with lupus nephritis (LN). Although persistent tubulointerstitial inflammation (TII) usually precedes tubulointerstitial damage (TID) in LN, the two conditions can be simultaneously present to varying degrees.

Methods: We examined whether coexisting TII/TID predicts progression to chronic kidney disease (CKD) in LN patients.

Results: Of the 175 LN patients enrolled in the study, 53 (30.2%) had coexisting TII/TID. Compared with LN patients without coexisting TII/TID, LN patients with coexisting TII/TID more often had LN of the proliferative type and a larger eGFR (98.5% vs 78.6%, p<0.001) and reaching LLDAS (HR 0.23, 95% CI 0.08-0.67, p=0.007) were both independently associated with a lower mortality.

Conclusion: This is the largest multicenter prospective SLE-PAH cohort to describe disease characteristics and prognosis. This study showed that survival has improved significantly for SLE-PAH. Early detection of PAH in SLE patients, achieving PAH treatment goal as well as LLDAS for SLE contributed to survival improvement.

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