Objectives: To assess the frequency of accrual damage in antiphospholipid syndrome (APS) and to evaluate the association with different laboratory and clinical APS subsets.

Methods: Medical records of 274 patients, 231 (84.3%) female and 43 (15.7%) males with a mean (±SD) age at diagnosis of 37.8 (±11.5) years, followed prospectively from 1990 to 2021, were reviewed.

Results: Ninety-six (35%) presented pregnancy morbidity alone, 140 (51.1%) thrombosis alone and 38 (13.9%) both thrombosis and pregnancy morbidity. A single, double or triple antiphospholipid antibodies (aPL) positivity was registered, respectively in, 82 (29.9%), 78 (28.5%) and 114 (41.6%) of the 92 patients. Following a mean (±SD) follow up of 208.4 (±17.1) months, a total of 58 (21.2%) organ damage accrual was recorded. This included neurological damage in 19 (32.8%) patients, hemiparesis in 9, epilepsy in 7 and cognitive dysfunction/dementia in three cases, cardiac valvopathy in 4 (6.9%) patients of which 3/4 (75%) require valve replacement with mechanical valve in two cases and bio-prosthesis in one. Chronic heart failure was found in 4 (6.9%) patients, chronic renal failure in 15 (25.9%), amputation due to peripheral arterial thrombosis in 5 (8.6%), visual loss in 2 (3.4%), post thrombotic syndrome in 6 (10.3%), adrenal insufficiency in one (1.7%). Some of the patients present more than one organ dysfunction. Both thrombotic and thrombotic and pregnancy morbidity subsets were significantly associated with a higher rate of damage accrual compared with pregnancy morbidity alone, respectively p<0.0001 (OD 40.7; 95% CI: 6.9-418.8) and p<0.0001 (OD 61.9; 95% CI 10.5-659.7). Moreover, the presence of microangiopathy as well as the presence of both venous and arterial thrombosis were significantly associated with damage accrual, respectively (p<0.0001, OD 10.99; 95% CI 5.7-21-36) and (p=0.001). Regarding laboratory subsets, triple aPL positivity was significantly associated with a higher rate of damage accrual compared to single and double aPL, respectively p=0.0001 (OD 9.6; 95% CI: 3.7-23.5) and p<0.0001 (OD 4.8; 95% CI: 2.2-10.81). At the multivariate analysis only, microangiopathy was an independent risk factor for damage accrual (p=0.001).

Conclusion: Overall, our data show a higher frequency of damage accrual in APS patients. Microangiopathy was independent risk factor for damage accrual. These findings should be in mind when counselling APS patients and might help guide clinicians in therapeutic decision.

REFERENCES:
1018. doi:10.1136/annrheumdis-2013-204838

Acknowledgements: NIL.

Disclosure of Interests: None Declared.

DOI: 10.1136/annrheumdis-2023-eular.5952

PULMONARY INVOLVEMENT IN LUPUS IS ASSOCIATED WITH ENHANCED MORBIDITY: A MULTICENTRE STUDY

Keywords: Lungs, Systemic lupus erythematosus, Autoantibodies

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Background: Although several studies report a high incidence of pulmonary manifestations in SLE, lung involvement is often underestimated in SLE clinical assessment [1]. This is also mirrored by the absence of pulmonary manifestations other than pleurisy in the old and new classification criteria for SLE [2]. Moreover, limited evidence on the management of SLE-related pulmonary manifestations is available.

Objectives: To assess prevalence and clinical impact of the spectrum of SLE-related pulmonary manifestations and their association with patient autoantibody profiles in a large SLE cohort and to describe the effectiveness of different therapeutic approaches in distinct clinical settings.

Methods: Patients followed at the Lupus Clinics of ASST G. Pini-CTO and San Raffaele Hospital (Milan, Italy) were enrolled. Data regarding demographics, disease characteristics, autoantibody profile, pulmonary manifestations, damage accrual and treatment were collected. The following types of lung involvement were recorded: pleurisy, acute lupus pneumonitis, interstitial lung disease (ILD), alveolar haemorrhage, pulmonary embolism, arterial pulmonary hypertension and shrinking lung syndrome.

Results: Of the 471 SLE patients enrolled, we identified 78 patients (16.5%) displaying at least one pulmonary manifestation. Epidemiological data on each of the 471 SLE patients was the most complete set of pulmonary manifestations and manifested at disease onset in most cases (56%). Patient home environment (urban vs countryside) did not seem to impact the risk of developing lung disease. Damage accrual was relevant, as 2/3 of patients displayed at least 1 point increase in SLICC Damage Index (SDI) after the onset of lung involvement in comparison to baseline. All patients received at least one steroid course. Immunosuppressive treatment choices and efficacy differed among distinct manifestations: only half of the patients with pleurisy received immunosuppression, predominantly azathioprine, with 100% of improvement, while 80% of cases of ILD received immunosuppression, predominantly mycophenolate, with a 50% risk of non-response. By comparing demographics and clinical characteristics among cases and controls, we found a significantly lower median age at disease onset (p=0.002) and a higher frequency of male sex (18% vs 9%; p=0.07), joint involvement (p=0.002) and constitutional symptoms (p=0.02) in patients with lung involvement, while no differences were observed in the autoantibody profile, including anti-dsDNA and anti-ENA autoantibodies.

Conclusion: Our study confirms that, in addition to the known epidemiological burden of pleurisy, other types of pulmonary involvement can complicate the disease course and contribute to damage accrual. In particular, ILD can frequently occur and respond to immunosuppressants in only half cases. Consistent with the association of lung involvement with increased morbidity, higher-risk categories for severe disease such as males and subjects with early-onset SLE were more represented among patients with pulmonary manifestations.

REFERENCES:

Table 1. Demographic and disease characteristics of SLE patients with lung involvement

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Females, n (%)</th>
<th>Males, n (%)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at diagnosis (years): median (IQR)</td>
<td>29 (22–39)</td>
<td>41 (33–47)</td>
<td>0.012</td>
</tr>
<tr>
<td>Disease duration (years): mean ±SD</td>
<td>19 ±12</td>
<td>21 ±13</td>
<td>0.36</td>
</tr>
</tbody>
</table>

| Subjects with >0 and >1 point SDI increase from baseline: n (%) | 48 (61.5) | 26 (33) |

Lung involvement: Females, n (%) | 65 (83) | 5 (6)

Aortic aneurysm | 4 (5) | 3 (3)
Aortic dissection | 2 (2) | 2 (2)
Pulmonary embolism | 4 (5) | 5 (5)
Shrinking lung | 1 (1) | 1 (1)
Arterial and venous | 5 (6) | 0 (0)
Neurological damage | 1 (1) | 0 (0)

Acknowledgements: NIL.

Disclosure of Interests: Maria Gerosa: None declared, Giuseppe Alvisse Ramirez: None declared, Lorenzo Maria Argolini: None declared, Isabella Scotti: None declared, Carolina Artusi: None declared, Luca Moroni: None declared, Enrica Bozzolo: None declared, Maria Rosa Pellico: None declared, Ludovica Cavallo: None declared, Lorenzo Dagna: None declared, Roberto Caporal Speakers bureau: AbbVie, Amgen, BMS, Celltrion, Fresenius, Galapagos, Janssen, Lilly, Novartis, Pfizer, and UCB, Consultant of: AbbVie, Fresenius, Galapagos, Lilly, Novartis, Pfizer, and UCB.

DOI: 10.1136/annrheumdis-2023-eular.5952

BASELINE CHARACTERISTICS OF A LONGITUDINAL, MULTINATIONAL, MULTITECHNIC STUDY OF LUPUS PATIENTS, WITH OR WITHOUT LUPUS NEPHRITIS

Keywords: Registries, Autoantibodies, Systemic lupus erythematosus