SLE, Sjögren’s and APS - clinical aspects (other than treatment)

BILAG-2004 INDEX ACTIVE DISEASE PREDICTS DEVELOPMENT OF DAMAGE


Background: BILAG-2004 systems tally showed that persistent minimal disease was protective of new damage (RR 0.99 with 95% CI: 0.99, 0.99) but not cumulative drug exposure (RR 2004 was predictive of development of damage in an inception cohort.

Methods: This was a prospective multi-centre longitudinal study in the UK of an inception cohort of SLE patients (recruited within 12 months of achieving 1997 ACR revised criteria for SLE). Data were collected on disease activity (BILAG-2004 and BILAG2004-Pregnancy Index during pregnancy), SLICC/ACR DI (SDI), cumulative drug exposure and death at each visit. Information on cardiovascular risk factors (hypertension, diabetes mellitus, hypercholesterolaemia and smoking status) and antiphospholipid syndrome status were also collected. This study ran from 1st January 2005 to 31st December 2017. Longitudinal analysis using Poisson regression with random effects model was used to determine predictors of development of new damage. Death was not included in the analysis due to small numbers.

Results: 273 patients were recruited (91.2% female, 59.3% Caucasian, 17.2% African/Caribbean, 17.2% South Asian with mean age at recruitment of 38.5 years (SD 14.8). 978% had no damage at recruitment (2.2% had SDI score of 1). Median follow-up was 73.4 months (range: 1.8, 153.8) with total follow-up of 1767 patient-years. Prevalence of risk factors during follow-up were: hypertension 23.1%, hypercholesterolaemia 35.5%, diabetes mellitus 5.5%, smoker or ex-smoker 44% and antiphospholipid syndrome 7%. There were 13 deaths and 114 new damage items occurred during follow-up.

We aimed to compare the performances of three criteria sets/rules in a large cohort of patients and relevant diseased controls from a reference center with dedicated clinics for SLE and other autoimmune/inflammatory connective tissue diseases from Turkey.

Methods: We reviewed the medical records of SLE patients and diseased controls for clinical and laboratory features relevant to all sets of criteria. Criteria sets/rules were analysed based on sensitivity, positive predictive value, specificity and negative predictive value, using clinical diagnosis with at least 6 months of follow-up as the gold standard. A subgroup analysis was performed in ANA positive patients for both SLE patients and diseased controls. SLE patients that did not fulfill 2012 SLICC criteria and 2019 EULAR/ACR criteria and diseased controls that fulfilled these criteria were evaluated.

Results: A total of 392 SLE patients and 45 controls met the criteria for the single center cohort from Turkey.

Conclusions: Active disease (Grade A or B) according to BILAG-2004 index is predictive of development of new damage in SLE patients.

References:

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vasculitis) were included into the study. Hundred and fourteen patients (16.6\%) were ANA negative. Sensitivity was more than 90\% for 2012 SLICC criteria and 2019 EULAR/ACR criteria and positive predictive value was more than 90\% for all three criteria (Table 1). Specificity was the highest for 1997 ACR criteria. Negative predictive value was 76.9\% for ACR criteria, 88.4\% for SLICC criteria and 91.7\% for EULAR/ACR criteria. In only ANA positive patients, sensitivity was 79.6\% for 1997 ACR criteria, 92.2\% for 2012 SLICC criteria and 96.1\% for 2019 EULAR/ACR criteria. Specificity was 92.6\% for ACR criteria, 87.8\% for SLICC criteria 85.2\% for EULAR/ACR criteria. Eleven clinically diagnosed SLE patients had insufficient number of items for both 2012 SLICC and 2019 EULAR/ACR criteria. Both criteria were fulfilled by 16 diseased controls; 9 with Sjögren's syndrome, 5 with antiphospholipid syndrome, one with dermatomyositis and one with systemic sclerosis.

### Table 1. Sensitivity, positive predictive value, specificity and negative predictive value of 1997 ACR, 2012 SLICC and 2019 EULAR/ACR classification criteria

<table>
<thead>
<tr>
<th></th>
<th>SLE (%)</th>
<th>Positive Predictive Value (%)</th>
<th>Value (%)</th>
<th>Specificity (%)</th>
<th>Negative Predictive Value (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1997 ACR</td>
<td>(+) 84</td>
<td>78.6</td>
<td>94.4</td>
<td>94.9</td>
<td>76.9</td>
</tr>
<tr>
<td>2012 SLICC</td>
<td>(+) 358</td>
<td>91.1</td>
<td>93.2</td>
<td>91.2</td>
<td>88.4</td>
</tr>
<tr>
<td>2019 EULAR/ACR</td>
<td>(+) 368</td>
<td>89.3</td>
<td>92.9</td>
<td>90.5</td>
<td>91.7</td>
</tr>
<tr>
<td>ACR</td>
<td>(+) 24</td>
<td>85.1</td>
<td>91.2</td>
<td>92.1</td>
<td>85.1</td>
</tr>
</tbody>
</table>

**Conclusion:** In this cohort, although all three criteria have sufficient specificity, sensitivity and negative predictive value of 1997 ACR criteria are the lowest. Overall, 2019 EULAR/ACR and 2012 SLICC criteria have a comparable performance, but if only ANA positive cases and controls are analysed, the specificity of both criteria decrease to less than 90\%. Some SLE patients with a clinical diagnosis lacked sufficient number of criteria. Mostly, patients with Sjögren's syndrome or antiphospholipid syndrome are prone to misclassification by both recent criteria.

**REFERENCES:**


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**POSO708 PSYCHIATRIC DISORDERS IN PATIENTS WITH DIFFERENT PHENOTYPES OF NEUROPSYCHIATRIC SYSTEMIC LUPUS ERYTHEMATOSUS (NPSLE)**

N. Monahan1, A. Blonk2, H. Middelkoop3, M. Kloppeburg4, T. Huizinga5, N. Van der Wee6, G. M. Steup-Beekman1, 7, 1Leiden University Medical Center, Department of Rheumatology, Leiden, Netherlands; 2Leiden University Medical Center, Department of Psychiatry, Leiden, Netherlands; 3Leiden University Medical Center, Department of Neurology, Leiden, Netherlands; 4Leiden University Medical Center, Department of Clinical Epidemiology, Leiden, Netherlands

**Background:** Patients with systemic lupus erythematosus (SLE) may present with psychiatric disorders. These are important to recognize, as they influence quality of life and treatment outcomes and strategies.

**Objectives:** We aimed to study the frequency of psychiatric morbidity as classified by the Diagnostic and Statistical Manual of Mental Disorders (DSM-V) in patients with SLE and neuropsychiatric syndromes of different origins.

**Methods:** In the neuropsychiatric SLE (NPSLE) clinic of the Leiden University Medical Center, patients undergo a standardized multidisciplinary assessment by a neurologist, neuropsychologist, vascular internal medicine, rheumatologist, physician assistant and psychiatrist. After two weeks, a multidisciplinary consensus meeting takes place, in which the symptoms are attributed to SLE requiring treatment (major NPSLE) or to minor involvement of SLE or other causes (minor/non-NPSLE). Consecutive patients visiting the NPSLE clinic between 2007-2019 were included. Data of psychiatric evaluation and current medication use were extracted from medical records. The presence of cognitive dysfunction was established during formal neuropsychological assessment.

**RESULTS:** 371 consecutive SLE patients were included, of which 110 patients had major NPSLE (30\%) and 41 patients had minor NPSLE (11\%). Mean age at diagnosis was 44 years old and 87% was female. The most frequently diagnosed psychiatric disorders in the total group were cognitive dysfunction (42\%) and depression (23\%), as shown in Table 1. Furthermore, anxiety was present in 5\% and psychotic disorders in 4\% of patients. In patients with minor/non-NPSLE, especially depression (26\% vs 15\%) and anxiety (6\% vs 2\%) were more common than in major NPSLE. Cognitive dysfunction (54\% vs 36\%) and psychotic disorders (6\% vs 4\%) were more common in patients with major NPSLE than minor/non-NPSLE.

Psychiatric medication was used in 33\% of patients, of which antidepressants and benzodiazepines are the most frequently (both: 18\% in both subgroups). Antipsychotics were more often used in patients with NPSLE (10\% vs 7\%) and benzodiazepines more often in minor/non-NPSLE (20\% vs 14\%). In addition, 17 patients (5\%) had a history of suicide attempt, which was more common in patients with major NPSLE than minor NPSLE (6\% vs 2\%).

**Conclusion:** Psychiatric morbidity, especially cognitive dysfunction and depression, are common in patients with lupus and differ between underlying cause of the neuropsychiatric symptoms (minor/non-NPSLE vs major NPSLE).