Figure 1. Pearson correlation between FS and time interval until lymphoma diagnosis

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OP0292

SURVEY ON THE PERCEPTIONS AND EXPERIENCES OF SPANISH LUPUS PATIENTS. RESULTS ABOUT KNOWLEDGE OF THE DISEASE AND RELATIONSHIP TO DISEASE-FELUPUS SURVEY

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Background: Despite great advances in the diagnosis and treatment of lupus, the scientific community does not know the perception of our patients regarding the knowledge of the disease and the relationship between patients and disease. Fatigue had the greatest impact on activities of daily living, yet the majority reported receiving no support or poor support in managing it¹.

Objectives: Through this initiative, it is intended to investigate about the knowledge of the disease and impact of the disease on quality of life of Lupus patients. Another objective is to give visibility to the current needs of people living with lupus.

Methods: It was performed a national survey with 1,263 interviews with Lupus patients who reside in Spanish territory and belonging to lupus patient associations in Spain. The survey was carried out by the Spanish Lupus Federation (FELUPUS) in collaboration with GSK company. Online interview lasting approximately 25 minutes. The collection of information was anonymously carried out from May 21 to June 30, 2020.

Results: 1.263 lupus patients were interviewed, 92% diagnosed with SLE and 8% with CLE. Survey sample is representative of the Lupus patient population in Spain [associated sampling error: 2.76%]. Questions about knowledge of the disease showed that 73% of patients considered that there is very little knowledge of the disease by society. Patient awareness of lupus increases as the disease progresses, so at the time of diagnosis, level of knowledge of the patient about Lupus is low in 92% of patients and at the moment of survey, 68% of patient had high knowledge. In terms of the perception of his illness, the affectionation of the kidneys and heart (97%), fatigue and skin rashes (97%), are the statements that generate the greatest consensus. The survey about the relationship to disease demonstrated that 3 out of 4 patients have symptoms related to the disease, muscle and joint pain (75%) and fatigue (74%) are the symptoms that cause the greatest discomfort (Graph 1). Remarkable degree of agreement with the fact of not being able to sunbathe (78%), as well as the lack of energy (61%) and weakness in the body (60%). Flare-ups (86%), followed by fatigue (78%) and pain (77%) cause great concern. At diagnosis, 92% of patients have some organic involvement and regarding the diagnosis, at present, a greater number of patients present damage to the CNS (17%) and bones (21%). Many patients do not understand the concept of organ damage, wrongly relating it to fatigue (38%) or joint pain (47%).

Graph 1.

Conclusion: Among the conclusions of the survey, it stands out that society and the general population are unaware of what lupus is, while in the case of lupus patients, knowledge increases as the disease progresses. Citizen awareness campaigns about this disease are necessary, where patient associations together with health authorities have a crucial job. On the other hand, 92% of patients present organ damage at diagnosis. This means that we are arriving late to the diagnosis of many patients, which makes it necessary to promote a close collaboration between Primary Care and Hospitals, to refer patients as soon as they suspect SLE.

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What worries the most to Lupus patients?

Question P12. Please indicate your level of concern with the following aspects of Lupus. Percentage of patients who have scored a 4 or 5 for each item (% T2B).

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OP0293

OPTIMIZING A TOOL TO IDENTIFY LUPUS FLARES IN DAILY CLINICAL PRACTICE: SLE-DAS FLARE VERSUS SELENA FLARE INDEX

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Background: In the management of patients with Systemic Lupus Erythematosus (SLE), it is of utmost importance to accurately identify lupus flares. There is a conceptual consensus definition of lupus flares [1]; however, the instruments used to identify flares in clinical trials, such as the SELENA Flare Index (SFI) are too cumbersome to apply in daily clinical practice. The SLE disease activity score (SLE-DAS) is a validated continuous measure of disease activity with higher sensitivity to change and validity in predicting damage accrual when compared

to SLEDAI-2K. SLE-DAS is quickly scored with its online calculator. An increase in SLE-DAS ≥ 1.72 was validated as a clinically meaningful worsening of SLE disease activity. [2]

Objectives: To compare the performance of SLE-DAS, classic SFI (c-SFI), revised SFI (r-SFI) and SLEDAI-2K in the identification of lupus flares in a real-life clinical setting.

Methods: We included patients with SLE fulfilling classification criteria [ACR (1997) and/or SLICC and/or EULAR/ACR], followed at an academic lupus clinic from January 2017 to June 2020, and presenting with lupus low disease activity score (LLDAS) at baseline.

Flares occurring after baseline were identified as fulfillment of the conceptual definition of flare, as assessed by the senior lupus expert at time of each outpatient visit. For each flare event, we evaluated the fulfillment of flare criteria according to c-SFI, r-SFI, SLEDAI-2K (score increase ≥ 4 points from baseline), and SLE-DAS (score increase ≥ 1.72 from baseline). As control visits without flare, we considered the first visit after baseline, where we assessed the four tools, excluding those where a flare was identified by the gold-standard expert evaluation. Sensitivity and specificity of the four flare tools were estimated and McNemar's test applied to assess differences with the gold-standard flare definition. The inter-instrument agreement with the gold-standard was assessed through Cohen's Kappa.

Results: We included 297 patients (female: 86.2%; mean age: 48.9 ± 14.6 years, mean disease duration: 12.5 ± 9.0 years). At baseline, all patients were in LLDAS, receiving ongoing antimalarials, immunosuppressants, and/or glucocorticoids in 91.0%, 43.8% and 33.6%, respectively. During follow-up, 22.2% developed flares. The analysis included 92 flares [musculoskeletal (40.2%); renal (23.9%), mucocutaneous (18.5%), haematological (5.9%), serositis (3.3%); multisystemic (8.7%)], with increase or change of treatment in 80.4% of these episodes, and 292 visits without flare considered as control.

There was no statistically significant difference between either SLE-DAS flare or c-SFI and the gold-standard expert flare definition ($p=0.41$ and $p=0.82$, respectively), while r-SFI and SLEDAI-2K flare were different from the gold-standard (Table 1). There was a strong agreement between SLE-DAS flare, c-SFI, r-SFI and the expert definition (Cohen's kappa, Table 1).

Table 1. Performance of the flare tools for the gold-standard flare definition.

	Sensitivity (%)	Specificity (%)	McNemar's ^{§§}	Kappa [§]
SLE-DAS flare	90.1	95.0	n.s.	0.836
c-SFI flare	91.1	96.5	n.s.	0.869
r-SFI flare	93.4	92.9	$p<0.01$	0.820
SLEDAI-2K flare	51.6	98.6	$p<0.0001$	0.590

^{§§} McNemar's test: significant difference from gold-standard if $p<0.05$; [§] Cohen's kappa agreement (0-1) with the gold-standard flare definition; n.s.: non-significant

Conclusion: The c-SFI and SLE-DAS showed the best performance in identifying SLE flares. The SLE-DAS flare definition is easier to apply and hence might be considered as an optimal tool to be used in daily clinical practice.

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OP0294

SJÖGREN'S SYNDROME ASSOCIATED LYMPHOMAS: CLINICAL DESCRIPTION AND 10-YEAR SURVIVAL

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Background: Sjögren's Syndrome (SS) is a chronic systemic autoimmune disease of unknown etiology, carrying the highest lymphoma risk among autoimmune diseases, with significant impact on mortality and morbidity of patients.

Objectives: To describe: i) the clinical phenotype of SS, ii) the histologic type, stage, treatment options regarding lymphomas and iii) the prognosis of patients with SS related lymphoproliferative disorders.

Methods: Eight hundred and fifteen consecutive SS patients' records from a single center fulfilling the 2016 ACR/EULAR were reviewed retrospectively for the purpose of this study. One hundred twenty-one patients with a diagnosis of non-Hodgkin Lymphoma (NHL) were identified and enrolled in the study population. Cumulative clinical, laboratory and histologic data were recorded and overall survival as well as event free survival curves were constructed using the Kaplan-Meier method. An event was defined as a disease progression, lymphoma relapse, treatment failure, histologic transformation, development of a 2nd lymphoma or death from any cause.

Results: From 121 pSS patients with lymphoma the most common histologic type encountered was MALT lymphoma (92/121, 76.0%) followed by DLBCL (11/121, 9.0%) and NMZL (8/119, 6.6%). The remaining 10 patients had various lymphomas of B (follicular, lymphoplasmacytic, chronic lymphocytic leukemia) and T cell origin (peripheral T cell lymphoma not otherwise specified, primary cutaneous T cell lymphoma, angioimmunoblastic t-cell lymphoma). Permanent salivary gland enlargement (66.1%, 80/121), palpable purpura (34.7%, 42/121), peripheral nervous involvement (9.9%, 12/121), interstitial lung disease (8.2%, 10/121) presence of serum cryoglobulins (38.7%, 43/111) and C4 hypocomplementemia (69.8%, 81/116) present at least 1 year before the development of lymphoma were the main pSS related features. The median age at lymphoma diagnosis was 58 years old (range 29-82) while MALT lymphomas developed earlier compared to DLBCL from pSS diagnosis (8 vs 3 OR= 3.84, 95%CI: 0.29 to 10.46; $p=0.0266$). The commonest biopsy proven extranodal sites included the labial minor salivary (43.8% patients) and parotid glands (30.5%) while 11% of patients had more than 1 extranodal sites affected. Bone marrow involvement was evident in 24.3% of patients (29/119) while nodal involvement in 35.5% (42/118). The majority of patients (65%) had limited disease (stage I or II). A watch and wait therapeutic policy was chosen in 40 patients while the rest received rituximab with or without chemotherapy. The 10-year survival and event free rates were 79% and 45.5% for MALT lymphomas, 40.9% and 24.2% for DLBCL and 46% and 31% for NMZL respectively (Figure 1). The Mantel-Cox log-rank comparison of the overall survival curves revealed a statistically significant difference ($p=0.0016$) among lymphoma subtypes.

Figure 1. Overall and event free survival of SS-associated lymphoma patients. A. Kaplan-Meier overall survival analysis. B. A Kaplan-Meier event free survival analysis.