AB0620

HIGH RISK OF MISTAKEN CLASSIFICATION OF PRIMARY ANTIPHOSPHOLIPID SYNDROME AS SYSTEMIC LUPUS ERYTHEMATOSUSACCORDING TO THE SLICC CRITERIA: ANALYSIS OF A COHORT OF 214 ANTIPHOSPHOLIPID PATIENTS

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Background: The diagnosis of systemic lupus erythematosus (SLE) is based on the association of clinical and biological manifestations and on clinical experience. In 2012, a major revision by the Systemic Lupus International Collaborating Clinics (SLICC) group sought to improve their sensitivity and specificity. In replications, the SLICC classification produced fewer errors than the previous version; its higher sensitivity but lower specificity meant that some patients could be classified with SLE although they had another disease. In fact, the distinction between PAPS, APS associated with SLE, and isolated SLE may be difficult in some cases because the two diseases share some clinical and biological manifestations.
Objectives: To assess the limitations of the SLICC (Systemic Lupus International Collaborating Clinics) classification criteria for systemic lupus erythematosus (SLE), in patients with primary antiphospholipid syndrome (PAPS).
Methods: Retrospective study of a cohort of APS patients (Sydney criteria). We successively excluded patients with[1] at least one "SLE-specific" manifestation (biopsy-proven SLE nephropathy, arthritis, cutaneous, or neurologic SLE manifestations, pericarditis, autoimmune haemolytic anaemia, oral and nasal ulcers, non-scarring alopecia, anti-dsDNA, and anti-Sm antibodies),[2] any other autoimmune connective tissue disease, and/or[3] antinuclear antibodies>1/320. Careful file review confirmed PAPS among the remaining patients. We then assessed the number of SLICC criteria each patient met.
Results: Among these 214 APS patients, we excluded 85 with at least one SLE-specific manifestation, 8 with another connective tissue disease, and 21 with antinuclear antibodies>1/320, leaving 100 patients with primary APS. Among them, 28% met at least 4 SLICC classification criteria including one clinical and one immunological criterion (antiphospholipid antibodies, aPL, by definition) and could thus theoretically be classified with SLE. Fourteen had an arterial phenotype (50%), 9 a history of catastrophic APS (32%), and 18 a triple-positive profile for aPL (64%). None had developed SLE during a median follow-up of 12 [6.5–17] years.
Conclusions: Because 28% of our patients with longstanding and strictly defined PAPS could be mistakenly classified as SLE, they were at risk of deleterious therapeutic management. We therefore suggest that any future classification for SLE should specifically require at least one SLE-specific criterion for patients with aPL.

REFERENCES:

[1] Petri M, Orbai A-M, Alarcón GS, et al. Derivation and validation of the Systemic Lupus International Collaborating Clinics classification criteria for systemic lupus erythematosus. Arthritis Rheum 2012;64:2677–86. doi:10.1002/art.34473
[2] Piette JC, Wechsler B, Frances C, *et al.* Exclusion criteria for primary antiphospholipid syndrome. *J Rheumatol*1993;**20**:1802–4.
[3] Asherson RA, Khamashta MA, Ordi-Ros J, *et al.* The 'primary' antiphospholipid syndrome: major clinical and serological features. *Medicine (Baltimore)* 1989;**68**:366–74.
[4] Gómez-Puerta JA, Martín H, Amigo M-C, *et al.* Long-term follow-up in 128 patients with primary antiphospholipid syndrome: do they develop lupus? *Medicine (Baltimore)* 2005;**84**:225–30.

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AB0621

EPIDEMIOLOGY, CLINICAL CHARACTERISTICS AND THERAPY APPORACHES OF A RETROSPECTIVE COHORT OF PAEDIATRIC SYSTEMIC LUPUS ERYTHEMATOSUS IN A TERTIARY CENTRE

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Background: Systemic lupus erythematosus (SLE) is a chronic autoimmune disease, potentially severe, with broad clinical spectrum, which can affect multiple organs and systems.
Objectives: To analyse the initial manifestations, laboratory examinations and therapeutic approaches of patients diagnosed with childhood onset systemic lupus erythematosus (c-SLE) followed in a tertiary hospital in the last 14 years.
Methods: Retrospective chart review. Inclusion criteria were: children under 18 years diagnosed with c-SLE between January 2003 and January 2017.
Results: During the study period, 38 patients were identified (ratio female/male: 3/1). The mean age at the onset of disease was 11.5 years (range 6–17). All had caucasic origin, except 5 coming from South America, 2 from India and 1 from Africa. Onset of the disease took place in spring in 22 (58%) patients, while in 10 (26%) patients onset was in summer, 5 (13%) in autumn and only one (3%) in winter season.
The most frequently clinical manifestations found at the debut were cutaneous involvement (66%, predominantly in the form of a malar rash), renal (65%) and joint (60%, 48% arthralgia and 52% polyarthritis). Other manifestations were fever (50%), cytopenias (39%), asthenia (36%), serositis and neurological clinic (both 26%) and oral aphthosis (23%). Among the neurological manifestations, 4 patients showed bradypsychia, 3 headache, 2 seizures and 1 manifested a paralysis of the sixth cranial nerve. Only 1 patient presented macrophage activation syndrome after primoinfection by *Ebstein Barr Virus*. During the study period, no deaths ocurred.
Ten patients also had some other associated autoimmune disease: 6 hypothyroidisms, 2 IgA deficiency, 1 vitiligo and 1 celiac disease. No case of diabetes mellitus was identified.
Regarding immunology, in all cases, positive antinuclear antibodies (ANA) were detected, although with variable titers (1/80 – 1/5120). Of the 25 patients who presented renal disease at the onset, 19 associated Anti-dsDNA antibodies positive at the initial time of the determination. Although antibodies related to antiphospholipid syndrome (APS) (anticardiolipin, anti-2glycoprotein and lupus anticoagulant) were detected in 12 patients (32%), only two developed associated clinical manifestations (both deep vein thrombosis in the lower limbs).
Regarding treatment, all patients required corticosteroids. Therapy with acetylsalicylic acid was indicated in all patients with APS-associated immunology. Only 6 patients received treatment with corticosteroids and hydroxychloroquine exclusively. Nineteen (50%) patients initially received azathioprine therapy, being necessary to switch to mycophenolate mofetil for lack of response in eleven, receiving the last treatment 20 patients finally (52%). Eighteen patients (47%) received cyclophosphamide therapy, 16 of them as a consequence of their renal involvement. In addition, biologic therapy (rituximab and belimumab, respectively) was used in two multirefractory patients.
Conclusions: As widely already reported, SLE is a disease that affects predominantly women. Moreover, as it has been previously described in the literature the most frequently initial manifestations found in c-SLE are cutaneous, renal and articular. However, a large variability of onset symptoms exists, thus c-SLE should be ruled out in patients with multisystemic involvement
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AB0622

CHARACTERISTICS OF SYSTEMIC LUPUS ERYTHEMATOSUS PATIENTS IN MALTA; A POPULATION BASED CROSS-SECTIONAL COHORT STUDY

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Background: Systemic Lupus Erythematosus (SLE) is an autoimmune disorder that involves multiple systems including the skin, musculoskeletal, renal, neurological, haematologic, cardiovascular and respiratory systems.
Objectives: The aim of this study was to characterise the patients with systemic lupus erythematosus living in Malta, in terms of age of disease onset, BMI, comorbidities, drug history, disease activity, damage and other factors including fatigue, sleep quality, depression, anxiety and vitamin D level.
Methods: The study consisted of a cross-sectional cohort study of all known SLE patients, over the age of 18, living in Malta. 92 patients who fulfilled the SLICC classification criteria for SLE, gave informed consent and an interview was carried out. Fatigue, anxiety, depression, sleep quality and disability were assessed respectively by filling in the following questionnaires: Fatigue Severity Scale (FSS), Hospital Anxiety and Depression Scale (HADS), Pittsburgh Sleep Quality Index (PSQI) and modified Health Assessment Questionnaire (mHAQ).
Results: 92.4% of SLE patients studied were female. Table 1 summarises the characteristics of the SLE patients.
23.9% of SLE patients were in remission (SLEDAI-2K 0), while 52.2% had a low disease activity (SLEDAI-2K 1–5) at the time of the interview. 20.7% and 3.3% had a moderate (SLEDAI-2K 6–10) and high (SLEDAI-2K 11–19) disease activity respectively. A significant positive correlation was noted between function measured by mHAQ and SLEDAI (R=0.417, p=0.000). 56.5% were noted to have an abnormally high level of fatigue (FSS >3.7). 6.5% were noted to have depression (HADS D>10) and 35.9% had anxiety (HADS A>10). 55.4% were noted to have poor sleep quality (PSQI >5) and 26.1% had an abnormal level of function (mHAQ >0.3). 15.2% were found to have vitamin D deficiency and 27.2% were vitamin D insufficient.

Abstract AB0622 – Table 1. Clinical characteristics of the cohort

Characteristics	Values
Age, mean (S.D.) years	46.9 (13.9)
Caucasian race, n/N (%)	90/92 (97.8)
Disease duration, median (range) years	13 (0–35)
Age of SLE onset, mean (S.D.) years	33.8 (12.8)
BMI, median (range) kg/m^2	26.5 (17.7–53.5)
Current smoker, n/N (%)	14/92 (15.2)
Family history of SLE in first degree relative, n/N (%)	3/92 (3.3)
Osteopaenia/osteoporosis, n/N (%)	30/92 (32.6)
Hypertension, n/N (%)	22/92 (23.9)
Diabetes mellitus, n/N (%)	7/92 (7.6)
Fibromyalgia, n/N (%)	9/92 (9.8)
Anti-phospholipid syndrome, n/N (%)	7/92 (7.6)
Sjogren's syndrome, n/N (%)	4/92 (4.3)
Rheumatoid arthritis, n/N (%)	3/92 (3.3)
Current prednisolone, n/N (%)	41/92 (44.6)
Current hydroxychloroquine, n/N (%)	55/92 (59.8)
Current azathioprine, n/N (%)	20/92 (21.7)
Current methotrexate, n/N (%)	10/92 (10.9)
Current mycophenolate, n/N (%)	6/92 (6.5)

Conclusions: This is the first population based study on SLE to be carried out in Malta. The prevalence of SLE in Malta is estimated to be 25.5 patients per 1 00 000 and the estimated incidence is 1.05 patients per 1 00 000 per year. A high frequency of obesity and vitamin D deficiency and insufficiency were noted in SLE patients. Other unmet needs include an uncontrolled disease activity, fatigue, poor sleep quality and anxiety.
Disclosure of Interest: None declared
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AB0623 PREGNANCY OUTCOME IN SYSTEMIC LUPUS ERYTHEMATOSUS: A RETORSPECTIVE STUDY

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Background: Pregnancy represents a challenge for patients with systemic lupus erythematosus. One of the major risk is the occurence of a flare during pregnancy. The influences are mutual, and the risk of complications depend mostly on the disease activity in the last 6–12 months before pregnancy. Therefore, these patients need a multidisciplinary approach, the obstetrician should collaborate with the rheumatologist and nephorologist.
Objectives: To determinate the associations between disease activity and pregnancy outcomes, and the risk factors that predict pregnancy complications and flare.
Methods: We present a retrospective study conducted between january 2010 and december 2015. We enrolled 35 pregnant patients, diagnosed with SLE with ages between 21 and 46 years old. All patients were followed up since the beginning of the pregnancy until delivery. We tested the correlations between different biomarkers and clinical manifestations of disease activity and pregnancy outcomes.
Results: Maternal complications occured in 71,24% of the cases. The most common complications were: miscarriages, hematologic abnormalities (anemias, thrombocytopenia), premature birth and preeclampsia. The prematurity rate was 11% and most common delivery mode was the caesarian section. Also, the planning of the pregnancy was a predictor of a good fetal outcome (p=0,01). The presence of the lupus anticoagulant was associated with prematurity (p=0,046) at univaryate analysis. Antiphospholipidic syndrome was associated in 12% of the cases. Univaryate analysis did not show a correlation between the presence of the syndrome and any pregnancy outcome.

Abstract AB0623 – Table 1. Risk Estimate preeclampsia in patients with APS

	Value	95% Confidence Interval	
		Lower	Upper
Odds Ratio for Preeclampsia	6667	,607	73 195
For cohort APS=yes	2417	1104	5292
For cohort APS=no	,363	,065	2014
N of Valid Cases	33		

Conclusions: Pregnancy should be planned when the disease is in remission. Lupus nephritis, is an important risk factor for preeclampsia. Also, the presence of lupus anticoagulant is a risk factor for preeclampsia, and hematologic determinations during pregnancy. The risk factors for pregnancy complications were; secondary antiphospholipid syndrome, presence of lupus nephropaty, thrombocytopenia. our study showes that the exacerbations depend on the disease activity in the moment of conception.
Disclosure of Interest: None declared
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AB0624 DYSLIPIDEMIA IN SYSTEMIC LUPUS ERYTHEMATOSUS: CORRELATION WITH DISEASE SEVERITY AND CYTOKINES

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Objectives: The patients with systemic lupus erythematosus (SLE) are obviously at high risk of cardiovascular disease (CVD) and the relationships between disturbed lipid metabolism and lupus activity remain to be elucidated. We evaluated dyslipidemia in association with disease severity, organ involvement and cytokines in patients with SLE.
Methods: Outpatients with SLE (n=105) and healthy controls (n=75) were recruited in this study. The concentrations of plasma tumour necrosis factor receptors A(sTNFRA), tumour necrosis factor receptors B(sTNFRB) and adipokine angiopoietin-like 4 (ANGPTL4)were measured by ELISA. The clinic and laboratory data were collected from patient records using electronic data processing. The data collected included serum lipid (TG, TC, HDL, LDL, ApoA1, ApoB), renal function (proteinuria, albuminuria, creatinine, blood urea nitrogen, uric acid), liver function (Alanine transaminase [ALT], glutamic oxalacetic transaminase [AST], total protein, albumin, globulin), blood system (lymphocyte, white blood cell [WBC], platelet[PLT], haemoglobin[HB]).
Results: Compared with the healthy controls, the level of serum TG, TC, LDL, ApoB were significantly increased, while HDL and ApoA1 were decreased. The serum levels of LDL and ApoB were positively correlated to SLEDAI, while HDL and ApoA1 were negatively correlated to SLEDAI. The patients with lupus nephritis had more severe dyslipidemia. The blood TG, TC, LDL, ApoB levels were positively correlated to 24 hour proteinuria and serum creatinine, urea nitrogen, uric acid. The blood levels of TG, TC, LDL, ApoB were positively correlated to serum total protein, albumin, globulin. The serum level of ANGPTL4 was positively correlated to HDL and ApoA1 as well as sTNFRA and sTNFRB with TG
Conclusions: Dyslipidemia in SLE was significantly correlated with SLEDAI and kidney involvement. The circulated levels of sTNFRA, sTNFRB and ANGPTL4 were associated with dyslipidemia in SLE.
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AB0625 SYSTEMIC LUPUS ERYTHEMATOSUS IN EGYPTIAN COHORT OF PATIENTS: A MULTICENTER STUDY

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Background: Systemic lupus erythematosus (SLE) is a chronic systemic autoimmune disease that mainly affects females in the reproductive age.[1] The disease presents with a diverse spectrum of clinical and immunological manifestations which has been studied in many countries and ethnic groups. Data from North Africa especially Egypt are minimal.
Objectives: To study the clinical and immunological characteristics of an Egyptian cohort with SLE and compare it with data from MENA region and international data.
Methods: In this retrospective study, data of 569 SLE patients who fulfilled the modified American College of Rheumatology (ACR) criteria for the diagnosis of SLE[2] were collected from three tertiary care centres in Cairo and Alexandria from the period of January 2014 to December 2017. Disease activity was assessed by using the SLE disease activity index (SLEDAI).[3]
Results: Of 569 patients 92.6%were females and 4.7% males with mean age at presentation 26.3±8.8 years and median disease duration four years (min 0.08-max 30 years). The main presenting symptom was musculoskeletal (arthritis/arthralgia) in 44.1% followed by fever in 39.4% and nephritis in 14.2%. Renal affection was present in 374 patients (65.7%) and renal biopsy was done in 268 patients with the most common is class III and IV lupus nephritis {18.3% and 14.1% respectively} (table 1). Antinuclear antibodies (ANA) was positive in all patients and immunofluorescence pattern was done in 256 patients; homogenous