SAT0232 PERCEPTION OF THE DISEASE IN PATIENTS WITH EARLY SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: Systemic lupus erythematosus is an autoimmune disease with a major impact on patient's quality of life.
Objectives: To evaluate patient's attitude toward early disease and factors that influence it.
Methods: Performed case-control study included SLE patients that fulfilled SLICC, 2012 classification criteria. The research included two groups of patients: early SLE – 1st group (disease duration ≤24 months) and non-early SLE – 2nd group control (disease duration >24 months). The pattern of the disease activity was assessed by patient global assessment (PGA), Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI-2K) and Systemic Lupus Activity Measure (SLAM), for SLE activity, SLICC/ACR Damage Index (DI) for disease irreversible changes and SF-8 for the Quality of Life (QoL).
Results: A total of 101 SLE patients with 34 in the 1st group (early SLE) and 67 in the 2nd group (non-early SLE) was analyzed. The disease activity showed high disease activity in both groups by SLEDAI (7,02±4,16 and 6,26±4,43 points, p>0,05) and SLAM (7,47±4,40 and 7,31±4,10 points, p>0,05) such as (46,97±19,39 vs 47,98±22,41 points). The QoL was appreciated as low, by both components (mental and physical), in groups. The damage index was higher in the 2nd group (0,23±0,43 and 1,07±1,29, p<0,001), which can be explained by the development of irreversible changes with the increase of disease duration.
The PGA in early SLE was influenced by subjective symptoms contained in SLAM index (r=0,48, p<0,05), such as fatigue and depression, and the level of the quality of life (r=0,65, p<0,001). Meantime, PGA in patients with longer disease duration (>2 years), was influenced by the presence of organ damage by SLICC/ACR DI (0,23, p<0,05) and objective findings of the disease activity contained in SLEDAI (r=0,33, p<0,005) and SLAM (0,44, p<0,001).
Conclusion: The disease recognition in patients with early SLE was determined by subjective and psycho-emotional signs, while in patients with longer disease duration it was influenced by organ damage and complications.
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SAT0233 CHARACTERISTICS OF PRIMARY SJÖGREN'S SYNDROME INCLUDING ULTRASOUND FINDINGS OF THE SALIVARY GLANDS, ESSDAI AND ESSPRI

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Background: Studies have shown that salivary gland ultrasonography (SGUS) may have a potential value in the diagnosis of Sjogren's Syndrome (SS).
Knowledge of the association between ultrasonography findings, disease activity and damage, serologic markers and patient report outcome is limited.
Objectives: To investigate whether the results of SGUS are associated with disease manifestations and damage measured by doctor-reported activity score index (ESSDAI) and serologic markers. Furthermore to investigate the contribution of patient reported outcome measure (ESSPRI) in disease monitoring.
Methods: Patients registered at Odense University Hospital with the diagnosis primary SS were included in a Danish cohort. The patients were characterized using the ESSDAI, ESSPRI, serologic markers and SGUS-findings in submandibular and parotid glands. Schirmer's test and salivary test were performed for measurement of tear and salivary production.
SGUS was performed using a linear transducer, Siemens (ACUSON Sequoia Ultrasound System) on the two parotid and two submandibular glands. SGUS images was scored according to the OMERACT SS severity scoring system from 0 to 3, where 2 is moderate and 3 severe(1). A reliability study was performed in advance of the present study.
Spearman´s r correlation coefficient was used to assess correlation between scores.
Results: The cohort consisted of 48 Caucasian patients diagnosed with primary SS. Details on patient characteristics are shown in table 1.

Table 1.

Sex, n (%)	
Women	46 (95.8)
Age, mean (95%CI)	60 (57-62)
Smoking, n (%)	
Smoker	1 (2.1)
BMI, n (%)	
< 18.5	5 (10.4)
18.5 – 24.9	20 (41.7)
25.0 – 29.9	12 (25.0)
30.0 – 34.9	10 (20.8)
> 35.0	1 (2.1)
Serologic markers, n (%)	
SSa positive	33 (68.8)
SSb positive	22 (45.8)
ANA positive	38 (79.2)
Cryoglobulin positive	9 (18.8)
ESSPRI 0-10, mean (95%CI)	
Dryness	7.3 (6.7-7.9)
Fatigue	7.1 (6.4-7.7)
Pain	5.9 (5.1-6.7)
SGUS, n (%)	
Score 0	6 (12.5)
Score 1	15 (31.3)
Score 2	13 (27.1)
Score 3	14 (29.2)
ESSDAI, n (%)	
ESSDAI < 5 (low-activity)	22 (45.8)
≤ 5 ESSDAI ≤ 13 (moderate-activity) ESSDAI ≥ 14 (high-activity)	17 (35.4) 9 (18.8)

The correlation between ESSDAI-scores and SGUS-scores was r = 0.153 (p = 0.299). The correlation between ESSDAI-scores and ESSPRI-scores (dryness, fatique, pain) was r = 0.071 (p = 0.632), r = 0.254 (p = 0.082) and r = -0.002 (p = 0.987). The correlation between SGUS-scores and ESSPRI-scores (dryness, fatique, pain) was r = 0.124 (p = 0.400), r = -0.292 (p = 0.044) and r = -0.459 (p = 0.001).

Conclusion: In a Danish cohort of SS most patients had SSa and ANA autoantibodies. SGUS demonstrated high damage (score 2-3) in approximately half of the patients. ESSDAI activity score did not correlate with SGUS damage scores or the ESSPRI. SGUS damage scores correlated with ESSPRI-scores of fatique and pain, but not dryness.
Associations between other factors of importance for damage and SGUS scores are to be analyzed. SGUS and the ESSPRI describe different SS-related dimensions and will probably contribute in disease monitoring in the future.
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SAT0234 SERUM AXL, FERRITIN, IGFBP4 AND STNFR2 AS BIOMARKERS OF PEDIATRIC SLE

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Background: Proteomic screening is an efficient approach for identifying protein biomarkers in various inflammatory diseases. Our preliminary proteomic analysis revealed elevated levels of serum Axl, Ferritin, IGFBP4 and sTNFR2 in adult patients with active lupus nephritis (LN) (1). However, the role of these serum biomarkers in pediatric systemic lupus erythematosus (SLE) patients has not been examined.
Objectives: To evaluate the performance of 4 serum protein markers for detecting disease activity in pediatric patients with SLE.
Methods: 83 pediatric patients who fulfilled ≥4 ACR criteria for SLE and 25 healthy controls were recruited for serological testing of 4 protein markers identified by antibody-coated microarray screen, namely Axl, ferritin, IGFBP4 and sTNFR2. SLE disease activity was assessed using the SLEDAI-2k score, renal disease activity was assessed by the renal SLEDAI (range 0-16; 0= inactive LN, ≥ 8= active renal). 57 patients had clinically active SLE (SLEDAI score ≥ 4 or having a flare) (28 active renal and 29 active non-renal SLE patients). In active renal

patients, concurrent renal biopsy was performed, unless contraindicated. The ISN/RPS criteria were used to assess the histopathologic features of LN. Those Patients were further subcategorized into 2 groups; active proliferative (ISN/RPS classes III/IV) and non-proliferative (classes I/II/V).

Results: The serum concentrations of Axl and ferritin were significantly higher in patients with active SLE than inactive SLE (3765±235 vs. 2513±130 pg/ml, *P* = 0.001) and (111±26 vs. 18±4 ng/ml, *P* = 0.0001) respectively. Serum Axl levels were significantly higher in active renal versus active non-renal SLE patients (3765±235.3 vs. 2825±200.7 pg/ml, *P* = 0.04). In the active renal patients with paired kidney tissue and blood samples, none of the biomarkers tested discriminated classes of LN, although serum Axl, ferritin and IGBPB4 levels were higher in the proliferative subgroup. The levels of Axl, ferritin and IGFBP4 correlated significantly with SLEDAI scores (Axl, r= 0.58, *P* <0.0001; ferritin, r= 0.53, *P* <0.0001; IGFBP4, r= 0.229, *P* = 0.03). However, only serum Axl levels correlated significantly with the renal SLEDAI (r= 0.46, *P* = 0.01). The levels of Axl, IFGBP4 and sTNFR2 correlated with decreased C3 levels (r= - 0.54, *P* <0.0001; r= - 0.29, *P*= 0.007; r= - 0.29, *P*= 0.007) respectively. Only serum Axl and ferritin correlated with urinary PCR (r= 0.42, *P* <0.0001; r= 0.22, *P*= 0.04) respectively. These markers were more specific, but less sensitive, in detecting concurrent SLE activity than elevated anti-dsDNA or decreased C3. The specificity values of serum ferritin and IGFBP4 for concurrent active lupus nephritis were higher than anti-dsDNA or C3. Serum ferritin was the best predictor of global SLE activity (AUC 0.81, *P* <0.0001), followed by C3 (AUC 0.79, *P* <0.0001) then Axl (AUC 0.71, *P*= 0.002), while both Axl and C3 were the best predictors of lupus nephritis activity (AUC 0.72, both).

Conclusion: In pediatric SLE patients, serum ferritin and Axl perform better than traditional yardsticks in identifying disease activity, either global or renal. The performance of these serum markers should be explored further in a longitudinal cohort of pediatric SLE patients.

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SAT0235

THE EFFECT OF ANTIPHOSPHOLIPID ANTIBODIES ON APTT WAVEFORM PATTERNS

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Background: Patients with antiphospholipid antibody (aPL) are said to be at increased risk for thrombosis, however it is difficult to predict whether they will develop thrombosis. In recent years, it has been revealed that the characteristics of the second derivative curve of APTT waveform with aPL positive patient is biphasic changes[1,2]. As first step in predicting the risk of thrombosis, we sought to understand the effect of aPL on APTT waveform patterns.

Objectives: To analyze the characteristics of APTT waveforms according to the background diseases and the presence of aPL

Methods: Patients who underwent coagulation function tests from 2017 to 2019 were analyzed. A coagulation waveform (Clot waveform: CW) was drawn using a fully automatic coagulation time measuring device manufactured by Instrumentation Laboratory From the APTT waveform, the 1st derivative curve (DC) indicating the coagulation speed and the 2nd DC indicating the coagulation acceleration were depicted to measure the 1st DC height, 2nd DC peak 1 time, and 2nd DC peak 1 height (Figure1[2]). Patients were devided into CTD with aPL-negative patients (group A), aPL-positive patients with no prior thrombosis (group B), and antiphospholipid antibody syndrome (APS) (group C). Patients characteristics and aPL (anti-cardiolipin [CL] antibody IgM, anti-CL antibody IgG, anti-CLβ2GP1 complex antibody, LA-APTT, and LA- DRVVT) status were examind. A further analysis was performed according to the numbers of positive aPL. Comparison between the three groups were made by the one-way ANOVA method, with significant differences set as p-values <0.1. Factors with significant differences were analyzed by Steel-Dwass test. APTT waveforms was analyzed according to the numbers of positive aPL by least squares methods. Furthermore, to determine the cut off value of APTT, 1st DC height, 2nd DC peak 1 time, and 2nd DC peak 1 height for each case with 2 or more positive aPLs and 3 positive aPLs, area under the curve (AUC) of the receiver operating characteristic (ROC) curve, sensitivity and specificity were caliculated.

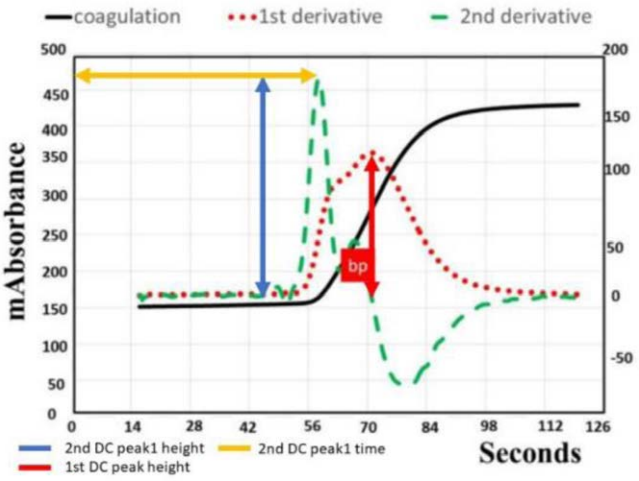

Figure 1.

Results: The APTT waveform was analyzed in 61 patients (51 women, 83.6%) with average age of 54.1 ± 17.1 years. Group A was 26 cases, Group B was 18 cases, and Group C was 17 cases. APTT, 2nd DC peak1 time, 2nd DC peak1 height, 1st DC peak time were significantly different among A, B, and C groups (p <0.01). APTT, 1st DC peak height, 2nd DC peak 1 time, and 2nd DC peak 1 height differed among the number of aPL (p < 0.01, respectively). APTT and 2nd DC peak1 time prolonged by 9.43 (seconds) and 16.3 (seconds) respectively according to the number of aPLs increased, and 1st DC peak height (mabs/s) and 2nd DC peak1 height (mabs/s^2) decreased by 56.4 (mabs/s) and 223.9 (mabs/s^2) respectively according to the number of aPLs decreased (Table 1). APTT> 35.2 (seconds) (sensitivity 80%, specificity 80.4%), 2nd DC peak1 height> 302 (mabs/s^2) (sensitivity 80%, specificity 91.3%) were relevant to the presence of two or more aPLs and APTT> 35.2 (seconds) (sensitivity 100%, specificity 80%), 2nd DC peak1 height> 302 (mabs/s^2) (sensitivity 100%, specificity 90%) were relevant to the presence of three aPLs.

Table 1.

The number of positive aPL	0	1	2	3	p value
The number of cases	27	19	4	11	
APTT	28.9	30.9	31.1	60.7	0.0001
(seconds)	[26.8, 31.4]	[29.1, 38.2]	[27.3, 54.2]	[45.9, 73.7]	
2nd DC peak time (seconds)	29.2	33.7	36.1	75.8	0.0001
	[26.8, 30.7]	[31, 41.7]	[31.5, 99]	[50.5, 102.4]	
2nd DC peak height (mabs/s^2)	839.9	669.6	608.4	119.3	0.0001
	[666.1, 962.2]	[346.4, 946]	[137.8, 956.7]	[30.6, 196]	
1st DC peak height (mabs/s)	309.6	271.5	241.8	135.6	0.0001
	[260.6, 355.1]	[168, 353]	[96.3, 364.0]	[76.8, 163]	

Conclusion: The presence of aPL was more related to the 2nd DC peak1 height of APTT waveform than APTT. A detailed review of the APTT waveform may further predict future thrombosis risk.

References:

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