

Abstract 127 Table 1 (Investigations)

Investigation	Case 1	Case 2	Case 3
Haemoglobin (g/L)	93	96	75
White cell counts (/L)	9.5×10 <sup>6</sup>	3.5×10 <sup>6</sup>	4.8×10 <sup>6</sup>
Lymphocyte count (/L)	1.9×10 <sup>6</sup>	1.2×10 <sup>6</sup>	0.3×10 <sup>6</sup>
Platelet count (/L)	145×10 <sup>9</sup>	147×10 <sup>9</sup>	37×10 <sup>9</sup>
Total serum bilirubin (mg/dl)	13.8		7.1
Conjugated bilirubin (mg/dl)	12		4
Aspartate aminotransferase (IU/L)	329	203	69
Alanine aminotransferase (IU/L)	96	156	57
Urine protein (mg/m <sup>2</sup> /hour)		24 mg/m <sup>2</sup> /hr	
C3 (Normal 50–150 mg/dL)	68	23	68
C4 (Normal 20–50 mg/dL)	4	4	3
ANA	3+diffuse	3+diffuse	4+speckled
Anti dsDNA (N:<60 IU/mL)	733	1621	689

presenting manifestation is uncommon in children with lupus.

**Methods** We retrospectively studied 140 pediatric lupus patients from January 1993- November 2018 and collected clinical and laboratory data of patients (3) who had liver involvement as presenting manifestation.

**Results** Case 1 was 13-year-old girl with fever and joint pain for 7 months associated with rash and yellow discoloration of eyes and body for 1 month. She also had altered behavior for 3 days. Examination revealed malar rash, pallor, jaundice, bilateral knee arthritis and hepatosplenomegaly. Investigations revealed (table 1) anemia, thrombocytopenia and lymphopenia. She had conjugated hyperbilirubinemia. Transaminases were elevated. Further investigations confirmed diagnosis of lupus (table 1). Markers for autoimmune hepatitis (SMA; LKM) were negative. She received pulse methylprednisolone followed by tapering doses of oral prednisolone. In view of neurological involvement she also received cyclophosphamide and shifted to azathioprine later. Her initial SLEDAI-2k was 22, that decreased to 4 at 2 year follow-up.

Case 2 was a 11-year-old boy with fever, rash and joint pain for 1 month. Examination revealed pallor, malar rash, oral ulcers, arthritis of bilateral elbow and knee and hepatomegaly. Investigations (table 1) revealed anemia; thrombocytopenia; leucopenia and lymphopenia. In view of multisystem involvement a possibility of lupus was considered which was confirmed by investigations (table 1). He had elevated transaminases. There was no coagulopathy. He was started on oral prednisolone. She also had class 3 lupus nephritis and received pulse cyclophosphamide followed by azathioprine in maintenance. Her transaminases decreased in follow-up. Her initial SLEDAI-2k was 18, that decreased to 2 at 2 year follow-up.

Case 3 was an 9-year-old girl who had rash and abdominal distention for 20 days and fever and altered sensorium for 6 days. On examination she had pallor, malar rash, ascitis and hepatosplenomegaly. Investigations revealed anemia; thrombocytopenia; leucopenia and lymphopenia. She had conjugated hyperbilirubinemia, elevated transaminases and coagulopathy. She also had deranged renal function tests. Ultrasonography reveals coarse echotexture, gross ascitis and splenomegaly. Possibility of lupus with acute on chronic liver decompensation was considered which was confirmed by

investigations (table 1). She was given methyl prednisolone. She had progressive azotemia so possibility of rapidly progressive renal failure was considered and cyclophosphamide was given. However, she developed refractory shock and eventually expired.

**Conclusions** Hepatic involvement at presentation in lupus can be multifaceted and poses challenge in diagnosis.

**Funding Source(s):** None

## 128 THE LUPUS SEVERITY INDEX IS A PREDICTOR OF DAMAGE AND DEATH IN LUPUS PATIENTS

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**Background** Predictors of poor outcome in systemic lupus erythematosus (SLE) may lead to the identification of high-risk patients at the onset of disease (incident cases) and/or when we first assess them in our clinics (prevalent cases). We tested whether the Lupus Severity Index (LSI) can help characterize high versus low risk lupus patients.

**Methods** Population: Patients from six lupus centers were recruited according to a standard data collection protocol. We characterized incident cases and prevalent cases as those with a diagnosis made within or after the previous 15 months. Data collected: Demographic, socioeconomic, disease specific and medication data were collected at baseline and annually. We collected: the American College of Rheumatology (ACR) and the Systemic Lupus International Collaborating Clinics (SLICC) classification criteria, the SLE Disease Activity Index (SLEDAI), the Systemic Lupus Activity Questionnaire (SLAQ), and the SLICC Damage Index (SDI). The LSI was derived from the ACR classification criteria and used as a predictor variable. Statistical analyses: Kruskal-Wallis test and Spearman correlations were used to see the association of LSI with categorical and continuous variables respectively. The baseline LSI was used to predict outcomes at follow-ups using logistic regressions and Spearman correlations for dichotomous and continuous variables respectively.

**Results** We enrolled 639 lupus patients and 440, 324 and 168 were re-evaluated at 1, 2 and 3 years. Baseline characteristics (table 1) [median (IQR)] were: age=49.0 (36.8–58.5) years, female=92%, Caucasian=74%, disease duration=10.1 (2.7–20.6) years. We had 129 (20%) incident cases and 471 (74%) prevalent cases with missing information in 39 (6%). Twelve patients died during follow-up. Table 1 summarizes baseline associations between LSI and several characteristics for the incident and prevalent cases. We found that age, sex, ethnicity (Asian worse LSI), SLICC classification criteria, SLEDAI, prednisone use and daily dose were associated with LSI in both incident and prevalent groups while the SDI and the use of immunosuppressors drugs was associated with LSI only in the prevalent cases. In follow-up, baseline LSI predicted SDI in prevalent cases (p=0.02) with a trend in incident cases (p=0.07). LSI predicted death in the prevalent group.

**Conclusions** The LSI is easy to derive from the ACR classification criteria and a useful measure of severity in lupus. The LSI is associated with baseline characteristics, some of them -

**Abstract 128 Table 1** Patients characteristics and associations with LSI by disease duration, at baseline

Variables *	Descriptive Statistics **	Association with LSI ***	
		Incident cases	Prevalent cases
<b>Patient's characteristics</b>			
Age	49.0 (36.8–58.5)	−0.19 (p=0.028)	−0.19 (p<0.001)
Gender		p=0.007	p=0.006
Women	589 (92)	5.4 (4.9–6.3)	6.3 (5.7–8.0)
Men	55 (9)	6.7 (5.6–8.3)	7.1 (5.8–8.5)
Ethnicity (n=592)		p=0.032	p=0.005
Caucasian	437 (74)	5.4 (4.7–6.4)	5.7 (5.1–7.9)
Asian	80 (14)	6.1 (5.4–8.5)	7.7 (5.3–8.2)
Other	75 (13)	5.9 (5.3–8.1)	5.9 (5.2–8.2)
Marital Status (n=586)		p=0.318	p=0.076
Single	107 (18)	5.7 (5.3–7.3)	6.5 (5.3–8.2)
Married	387 (66)	5.4 (4.7–6.3)	5.8 (5.2–8.0)
Other	92 (16)	5.4 (4.8–7.1)	5.7 (4.9–8.2)
Site		p=0.789	p=0.383
Winnipeg	231 (36)	5.8 (4.9–7.0)	5.6 (5.0–7.8)
Hamilton	25 (4)	6.2 (5.4–6.6)	6.6 (5.4–7.1)
Quebec	129 (20)	5.4 (4.6–6.8)	6.2 (5.2–8.3)
Ottawa	4 (1)	6.1 (6.1–6.1)	6.3 (5.7–8.2)
London	6 (1)	7.2 (7.2–7.2)	6.1 (5.5–7.8)
Calgary	244 (38)	5.4 (5.0–6.3)	5.9 (5.1–8.1)
<b>Disease's characteristics</b>			
Disease duration (n=623)	10.1 (2.7–20.6)	0.11 (p=0.241)	0.07 (p=0.106)
Recent diagnosis (<=15 months)		NC	NC
Ancient diagnosis (>15 months)			
ACR Criteria Score (n=626)	5 (4–6)	0.29 (p<0.001)	0.30 (p<0.001)
ACR LSI Score (n=626)	5.8 (5.1–7.9)	NC	NC
SLICC Criteria Score (n=538)	6 (5–8)	0.49 (p<0.001)	0.42 (p<0.001)
SLEDAI score (n=598)	2 (0–6)	0.05 (p=0.586)	−0.00 (p=0.992)
SLAQ Score (n=407)	12 (7–18)	−0.26 (p=0.023)	−0.18 (p=0.001)
SDI Score (n=609)	0 (0–2)	p=0.296	p=0.005
0	310 (51)	5.5 (4.8–6.4)	5.6 (4.9–7.9)
>0	299 (49)	5.5 (5.2–8.0)	6.2 (5.2–8.2)
<b>Medication</b>			
Prednisone (n=614)		p<0.001	p<0.001
Yes	203 (33)	6.4 (5.3–8.2)	6.8 (5.5–8.2)
No	411 (67)	5.4 (4.7–6.0)	5.6 (4.9–7.7)
Prednisone dose (n=219)	7.5 (5.0–15.0)	0.41 (p=0.001)	−0.01 (p=0.92)
Immunomodulators (n=616)		p=0.189	p=0.028
Yes	554 (90)	5.5 (5.0–6.9)	5.9 (5.2–8.1)
No	62 (10)	5.3 (4.4–6.2)	5.5 (4.9–6.4)
Antimalarial		p=0.241	p=0.97
Yes	517 (83)	5.5 (4.9–6.8)	5.8 (5.1–8.1)
No	106 (17)	5.2 (4.6–6.3)	5.8 (4.9–8.2)
Biologic Agent (n=609)		p=0.800	p=0.270
Yes	25 (4)	5.9 (5.3–6.5)	6.1 (5.3–8.4)
No	584 (96)	5.5 (4.9–6.8)	5.8 (5.1–8.1)
<b>Death during follow-up</b>			
		NC	p=0.002
Yes	12 (2)		8.2 (7.0–9.3)
No	627 (98)		5.8 (5.1–8.1)

\*n=639 except when specified; \*\*Results presented as Median (1 st quartile 3rd quartile) or Frequency (percentage) for continuous and categorical variables respectively; \*\*\*Results presented as Spearman Correlation coefficient (pvalue) or Median (1 st quartile 3rd quartile) with Kruskal-Wallis pvalue for continuous and categorical variables respectively; NC: Not calculated.

like disease activity and prednisone dose - modifiable. LSI can predict adverse outcome such as damage or death over time.

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### MACROPHAGE ACTIVATION SYNDROME AS A PRESENTATION IN PEDIATRIC LUPUS: A RETROSPECTIVE STUDY OF 3 CASES

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**Background** Macrophage activation syndrome (MAS) can, at times, be the presentation of pediatric lupus and diagnosis requires high index of suspicion.

**Methods** We retrospectively studied 140 pediatric lupus patients from January 1993- November 2018 and collected clinical and laboratory data of patients (3) who had MAS as presenting manifestation.

**Results** Case 1 was 11-year-old girl with fever for 4 months associated with rash and generalized body swelling for 1 month. Examination revealed rash over malar area and ear lobules, anasarca, hepatomegaly, bilateral pleural and pericardial effusion. In view of multisystem involvement a possibility of lupus was considered which was confirmed by investigations (table 1). She had elevated ferritin and fasting triglyceride and low fibrinogen. A clinical possibility of lupus with associated MAS was considered. She received pulses of methylprednisolone, one dose of intravenous immunoglobulin following which she improved. In view of nephrotic range proteinuria she was started on induction regimen with cyclophosphamide and shifted to mycophenolate in maintenance. Her initial SLEDAI-2k was 32- this decreased to 4 at 3 year follow-up.

Case 2 was a 9-year-old girl with fever, rash, generalized body swelling for 1 month and altered sensorium for 4 days. Examination revealed pallor, oral ulcers and hepatomegaly. She was in shock at presentation. In view of multisystem involvement a possibility of lupus was considered which was confirmed by investigations (table 1). She had pericardial effusion and low ejection fraction (25%). A possibility of MAS was considered and investigations revealed hyperferritinemia, elevated triglyceride and hypofibrinogenemia. She was given methylprednisolone pulses and continued on oral prednisolone, mycophenolate and hydroxychloroquine. Her initial SLEDAI-2k was 17- this decreased to 0 at 3 year follow-up.

Case 3 was an 8-year-old girl who had fever, rash and body swelling for 15 days. She had tachycardia, tachypnea, pallor, anasarca, subconjunctival bleed and frontal alopecia. She had pleural and pericardial effusion. In view of multisystem involvement a possibility of lupus was considered which was confirmed by investigations (table1). She had high ferritin and triglyceride. So a possibility of MAS was considered and she received pulse methyl prednisolone and intravenous immunoglobulin. She also had hematuria and proteinuria (renal biopsy could not be performed as she was sick and had thrombocytopenia) so was given pulse cyclophosphamide