



---

# Demographic Presentation, Activity Indices, Damage Index: Comparative Study Between Pediatric Lupus Erythematosus Versus Adult Systemic Lupus Erythematosus in Sample Egyptian Population

Eman Hassan Elsayed Hassan, Amira Hassan El-Gerby

Rheumatology Department, Faculty of Medicine, Alexandria University, Alexandria, Egypt

## Email address:

emanhassan96@yahoo.com (E. H. E. Hassan)

## To cite this article:

Eman Hassan Elsayed Hassan, Amira Hassan El-Gerby. Demographic Presentation, Activity Indices, Damage Index: Comparative Study Between Pediatric Lupus Erythematosus Versus Adult Systemic Lupus Erythematosus in Sample Egyptian Population. *American Journal of Internal Medicine*. Vol. 4, No. 1, 2016, pp. 12-18. doi: 10.11648/j.ajim.20160401.13

---

**Abstract:** Systemic lupus erythematosus (SLE) is a complex autoimmune disease that can affect all organ systems due to alterations of both the innate and adaptive immune systems. Although onset during infancy is rare, the incidence of SLE rises steadily during childhood until mid-adulthood, especially among females. In this study we aimed to highlight the possible discrepancies in clinical presentations as well as serological profiles of pediatric and adult onset SLE patients, we also focused attention on the disease assessment by SLE activity index (SLE DDI) and damage index at time of presentation. Subjects were subdivided into 2 groups: Group I: A total of 92 Pediatric systemic lupus erythematosus (pSLE) that were selected from the students attending the school children hospital of medical health insurance. Group II: A total of 90 adult systemic lupus erythematosus (aSLE) patients and were recruited from those attending the Alexandria Main University Hospital and outpatient clinic. All patients were subjected to: detailed history taking and complete physical and mental examination, also activity indices as well as damage index were applied for every lupus patient of the studied groups, laboratory investigations were done for all patients. Our results demonstrated that, regarding mucocutaneous manifestations: pSLE patients have values higher than aSLE patients regarding photosensitivity (63.3% and 61.1%) and vascular lesions (23.9% and 22.2%) respectively. Regarding haematological manifestations: pSLE patients have values higher than aSLE patients regarding anemia (86.96% and 84.4), leucopenia (28.3% and 22.22) and thrombocytopenia (46.7% and 25.56%) respectively. Regarding renal abnormalities, pSLE patients have higher incidence of nephritic syndrome than aSLE patients. Regarding SLEDAI, pSLE patients have values statistically higher than aSLE patients. Regarding SLAM, pSLE patients have values statistically higher than aSLE patients, while no differences of damage index was noticed.

**Keywords:** Demographic Presentation, Adult SLE, Pediatric SLE, Activity Indices, Damage Index

---

## 1. Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease affecting multiple organ systems triggered by the production of auto antibodies. [1]

SLE presents throughout the age spectrum. An estimated 10-20% of patients experience the onset of SLE prior to adulthood. More precise estimates are difficult due to a lack of a clear age limit for diagnosis of pediatric SLE. The maximum age at diagnosis most commonly used to define pediatric SLE is 16 years but ages ranged from 14-20 years in various studies. [2, 3]

Although there are limited studies directly comparing adult and childhood onset SLE, it has been suggested that pediatric lupus patients have a more aggressive disease course and an increased rate of more unusual clinical presentations compared with their adult counterparts. [4]

Delay in SLE diagnosis is associated with higher mortality and a reduced likelihood of achieving remission. [5]. In adult with SLE remission for 1-years is required in as many as 6.5% of the patients conversely, despite a lack of firm estimates, remission is exceedingly rare in pediatric SLE. [6]

Comparisons of relatively small pediatric and adult cohorts of SLE patients have shown that children and adolescents

have more active lupus, in particular lupus nephritis, at presentation and over time than adults. Compared to adults with lupus, children receive more intensive drug therapy and accrue more end-organ damage, often related to steroid toxicity [2]. In the Lupus in Minorities: Nature vs. nurture (LUMINA) multiethnic cohort, young age was an important independent predictor of new or worsening proteinuria on routine screening, and adolescent onset of SLE resulted in more aggressive disease and worse outcomes. [7, 8] The high morbidity and mortality observed from lupus nephritis in past studies of SLE in children may be due to delays in diagnosis and treatment [1, 3, 4].

Major cause of death in pediatric SLE and adult SLE include renal disease, severe disease flares, and infections. [7]

New psychiatric lupus is a risk factors of poor outcome in pediatric SLE, cardiovascular disease remains on important cause of death in a SLE.

There is a controversy as to whether age at SLE onset constitutes a risk factors for poor outcome. [8]

Despite improved survival rates in SLE patients of all age, there remains substantial morbidity due to disease damage.[7]

In a SLE, increasing age and larger duration of disease are correlated with disease damage. [8]. There is a trend towards higher rates of any disease damage in adolescent-onset SLE patients. [6]

In this study we aimed to highlight the possible discrepancies in clinical presentations as well as serological profiles of pediatric and adult onset SLE patients, we also focused attention on the disease assessment by SLE activity index (SLE DDI) and damage index at time of presentation.

## 2. Subject and Methods

This is a comparative study conducted between December 2013 till December 2015, included 182 patients fulfilling the systemic lupus international collaborating clinics (SLICC) 2012 criteria for diagnosis of SLE [9], patients were subdivided into 2 groups:

- *Group I:* A total of 92 Pediatric systemic lupus erythematosus (pSLE) patients that were selected from inpatients and outpatients clinic of students attending the sporting school children hospital of medical health insurance.
- *Group II:* A total of 90 adult systemic lupus erythematosus (aSLE) patients that were recruited from those attending the Alexandria Main University Hospital and outpatient clinic.

### 2.1. All Patients Were Subjected to

- I. Detailed history taking and complete physical and mental examination.
- II. Clinical assessment of activity including:
  - i. SLE Disease Activity Index (SLEDAI): [10] Minimum score is 0 and maximum is 105. (score less than 4 as inactive, from 4 to 8 as mild, from 9 to 12 as moderate and score more than 12 as having severe

disease activity).

- ii. Systemic lupus activity measure (SLAM). [11]. Which include subjective features reported by the patients, the higher the number, the more active the disease.
  - iii. Damage Index (SLEDDI): [12] Maximum possible score is 47.
- III. Laboratory investigations done for the studied group of patients included:
- i. Complete blood picture,
  - ii. liver enzymes (ALT, AST),
  - iii. renal function test (blood urea, serum creatinine, creatinine clearance and 24 hour urine proteins and urinary albumin creatine ratio).
  - iv. complete urine analysis,
  - v. erythrocyte sedimentation rate (ESR),
  - vi. C-reactive protein (CRP),
  - vii. C3, C4,
  - viii. lipid profile including serum cholesterol, triglycerides,
  - ix. antinuclear antibodies (ANA) titre.
  - x. antidouble stranded DNA antibodies (anti-ds DNA) titre.

### 2.2. Statistical Analysis of the Data

Data were fed to the computer and analyzed using IBM SPSS software package version 20.0. Comparison between different groups regarding categorical variables was tested using Chi-square test. Normally quantitative data was compared using student t-test, or F test (ANOVA), abnormally distributed data was compared using Mann Whitney test or Kruskal Wallis test, Correlations between two quantitative variables were assessed using Pearson or Spearman coefficient according to test of normality. Significance of the obtained results was judged at the level of 0.05.

## 3. Results

Table (1) shows the demographic data of the studied patients groups, it demonstrated that,

### *Age*

Age ranged from 8.0-16.0 and 21-52.0 years with the mean of 11.80±2.52 and 36.8±11.3 years for children and adult groups respectively, there were statistical significant differences between the two studied groups. (P=0.0001).

### *Sex*

This study include 8 (8.7%) and 11 (12.2%) males and 84 (91.3%) and 79 (87.8%) females for children and adult groups respectively with no statistical significant differences. (P=0.287).

### *Disease duration*

Disease duration ranged from 2.0-62.0 months and 4-58.0 months with the mean of 29.80±16.76 and 31.2±12.3 months for children and adult groups respectively with no statistical significant differences. (P=0.425).

### *Family history*

Positive family history was found in 5 (5.4%) and 6 (6.7%)

for children and adult groups respectively with no statistical significant differences. (P=0.725).

**Table 1.** Demographic data of the studied patients group.

	Group I (pSLE) "No. 92"		Group II (aSLE) "No. 90"		p
	No.	%	No.	%	
Age					
Range	8.0-16.0		21-52.0		
Mean	11.80		36.8		0.0001*
±S. D.	2.525		11.3		
Sex					
Males	8	8.7	11	12.2	0.287
Females	84	91.3	79	87.8	
Disease duration (months)					
Min	2.00		4.00		0.425
Max	62.00		58.0		
Mean	29.80		31.2		
± SD	16.76		12.3		
Family history					
Yes	5	5.4	6	6.7	0.725
No	87	94.6	74	82.2	

**Table 2.** Clinical data of the studied group of patients.

	Group I (pSLE) "No. 92"		Group II (aSLE) "No. 90"		p
	No.	%	No.	%	
Mucocutaneous					
Oral ulceration	9	9.78	20	22.2	
Photosensitivity	61	66.3	55	61.1	
Alopecia	30	32.6	32	35.6	
Discoid rash	2	2.17	5	5.56	
Livedo-reticularis	1	1.1	16	17.8	
Vascular lesions	22	23.9	20	22.2	
Articular complaints					
Myositis	3	3.30	12	13.33	
Arthralgia	80	87.00	40	44.44	
Arthritis	63	68.48	25	27.78	
Avascular necrosis of hip	1	1.1	3	3.3	
Constitutional manifestation					
Fever	24	26.1	25	27.8	
Fatigue, loss of weight	32	34.8	35	38.9	
Haematological					
Anemia	80	86.96	76	84.4	
Leucopenia	26	28.3	20	22.22	
Thrombocytopenia	43	46.7	23	25.56	
Ocular retinal changes	1	1.1	4	4.4	
Hypertension	18	19.6	28	31.11	
Cardiac					
Precardial effusion	7	7.61	11	12.22	
Pulmonary					
Pleural effusion	13	14.13	12	13.33	
Pulmonary Embolism	1	1.1	7	7.78	
Renal					
Nephritic syndrome	30	32.61	16	17.8	
Renal failure	2	2.2	3	3.3	
Neuroschiatric					
Seizure	2	2.2	3	3.3	
CVS	1	1.1	9	10	
Transvers myelitis	1	1.1	1	1.1	
Depression	7	7.6	33	36.67	
Headache	58	63.0	62	68.9	

### 3.1. Clinical Data

Table (2) shows clinical data of the studied patients groups, it illustrated that, Regarding mucocutaneous manifestations, pSLE patients have higher values than aSLE patients regarding photosensitivity (63.3% and 61.1%) and vascular lesions (23.9% and 22.2%) respectively. While, aSLE patients have values higher than pSLE patients regarding oral ulceration (22.2%, 9.78%), alopecia (35.6%, 32.6%), discoid rash (5.56% and 2.17%), livedo-reticularis (17.8%, 1.1%) respectively.

Regarding articular complaints, pSLE patients have higher values than aSLE patients regarding arthralgia (87.0% and 44.44%), arthritis (68.48% and 27.78%) respectively, while, aSLE patients have higher values than pSLE patients regarding myositis (13.33% and 3.30%), avascular necrosis of hip (3.3% and 1.1%) respectively.

Regarding constitutional manifestation, aSLE patients have higher values than pSLE patients regarding fever (27.8% and 26.1%), fatigue, loss of weight. (38.9% and 34.8%) respectively.

Regarding haematological manifestations, pSLE patients have higher values than aSLE patients regarding anemia (86.96% and 84.4), leucopenia (28.3% and 22.22) and thrombocytopenia (46.7% and 25.56%) respectively.

aSLE patients have values higher than pSLE patients regarding ocular retinal changes (4.4% and 1.1%), hypertension (31.11% and 19.6%), pericardial effusion (12.22% and 7.61) respectively.

Regarding pulmonary, pSLE patients have higher values than aSLE patients regarding pleural effusion (14.13 and 13.33%), while, aSLE patients have higher values than pSLE patients regarding pulmonary embolism (7.78% and 1.1%) respectively.

Regarding renal, pSLE patients have higher values than aSLE patients regarding nephritic syndrome (32.61 and 17.8%), while, aSLE have higher values than pSLE regarding renal failure (3.3% and 2.2%) respectively.

Regarding neuropsychiatric manifestations, aSLE patients have higher values than pSLE patients regarding seizure (3.3 and 2.2%), CVS (10% and 1.1%), depression (36.67% and 7.6%), headache (68.9% and 63.0%) respectively.

### 3.2. Complete Blood Picture

Table (3) shows complete blood picture of studied patients, it illustrated that,

#### Haemoglobin concentration (g/dl)

Haemoglobin concentration ranged from 6.0-12.80 and 6.5-14.1 with the mean of 9.59±1.91 and 10.11±2.01 for pSLE patients and aSLE patients respectively with no statistical significant differences (P=0.365)

#### WBCs count (x10<sup>3</sup> cell/mm<sup>3</sup>)

WBCs count ranged from 1.10-13.20 and 2.03-14.1 with the mean of 5.13±2.52 and 5.87±2.68 for group I and II respectively with no statistical significant differences (P=0.468).

#### Lymphocyte count (x10<sup>3</sup> cell/mm<sup>3</sup>)

Lymphocyte count ranged from 350-3000 and 365-3500 with the mean of 1800.7±640.1 and 1950.0±710.0 for pSLE patients and aSLE patients respectively with no statistical significant differences (P=0.225)

*Platelet count (x10<sup>3</sup> cell/mm<sup>3</sup>)*

Platelet count ranged from 35-410 and 40.0-450.0 with the mean of 205.1±77.8 and 218.6±81.6 for pSLE patients and aSLE patients respectively with no statistical significant differences (P=0.33)

**Table 3.** Complete blood picture of studied patients.

	Group I (pSLE) "No. 92"	Group II (aSLE) "No. 90"	P
Haemoglobin concentration (g/dl)			
Min	6.00	6.5	
Max	12.80	14.1	
Mean	9.59	10.11	0.365
± SD	1.91	2.01	
WBCs count (x10 <sup>3</sup> cell/mm <sup>3</sup> )			
Min	1.10	2.03	
Max	13.20	14.1	
Mean	5.13	5.87	0.468
± SD	2.51	2.68	
Lymphocyte count (x10 <sup>3</sup> cell/mm <sup>3</sup> )			
Min	350	365	
Max	3000	3500	
Mean	1800.7	1950.0	0.225
± SD	640.1	710.0	
Platelet count (x10 <sup>3</sup> cell/mm <sup>3</sup> )			
Min	35	40.0	
Max	410	450.0	
Mean	205.1	218.6	0.33
± SD	77.8	81.6	

**Table 4.** Routine investigations of studied patients.

	Group I (pSLE) "No. 92"	Group II (aSLE) "No. 90"	P
Fasting blood sugar (mg/dl)			
Min-Max	56-100	72-320	
Mean ± SD	86.2±17.0	182.5±45.1	0.002*
Serum cholesterol (mg/dl)			
Min-Max	61-260	95.0-310.0	
Mean ± SD	152.1±52.3	185.9±65.2	0.021*
Serum triglycerides (mg/dl)			
Min-Max	37-160	40.0-210.0	
Mean ± SD	80.7±48.2	131.2±52.6	0.003*
Blood urea (mg/dl)			
Min-Max	7-67	11-78	
Mean ± SD	32.1±20.6	36.5±16.8	0.221
Serum creatinine (mg/dl)			
Min-Max	0.4-4.1	0.94-4.96	
Mean ± SD	2.01±1.33	2.69±2.01	0.369
Creatinine clearance (ml/min)			
Min-Max	22-89	20.2-90.0	
Mean ± SD	62.1±21.4	52.6±24.3	0.112

	Group I (pSLE) "No. 92"	Group II (aSLE) "No. 90"	P
ALT (U/L)			
Min-Max	10-305	15-322	
Mean ± SD	45.2±50.2	56.9±45.8	0.098
AST (U/L)			
Min-Max	10-230	29.5-310.0	
Mean ± SD	43.7±39.6	58.0±42.1	0.211
ESR			
Min-Max	12-110	5-120	
Mean ± SD	47.33±18.77	61.33±27.11	0.016*
Urinary Alb/creatinine ratio			
Min-Max	22-105	20-122	
Mean ± SD	61.3±28.7	70.8±30.1	0.041*
CRP			
+ve	3	7	
-ve	89	83	0.098

### 3.3. Routine Investigations

Routine investigations of the studied patients were presented in table (4), it showed that

*Fasting blood sugar (mg/dl)*

Fasting blood sugar ranged from 56-100 and 72-320 with the mean of 86.2±17.0 and 92.5±22.6 for pSLE patients and aSLE respectively with no statistical significant differences (P=0.103).

*Serum cholesterol (mg/dl)*

Serum cholesterol ranged from 61-260 and 95.0-310.0 with the mean of 152.1±52.3 and 185.9±65.2 for pSLE patients and aSLE patients respectively with statistical significant differences, aSLE patients have statistically higher values than pSLE patients (P=0.021).

*Serum triglycerides (mg/dl)*

Serum triglycerides ranged from 37-160 and 40.0-210.0 with the mean of 80.7±48.2 and 131.2±52.6 for pSLE patients and aSLE patients respectively with statistical significant difference, aSLE patients have statistically higher values than pSLE patients (P=0.003).

*Blood urea (mg/dl)*

Blood urea ranged from 7-67 and 11-78 with the mean of 32.1±20.6 and 36.5±16.8 for pSLE patients and aSLE patients respectively with no statistical significant differences between the two studied groups (P=0.221).

*Serum creatinine (mg/dl)*

Serum creatinine ranged from 0.4-4.1 and 0.94-4.96 with the mean of 2.01±1.33 and 2.69±2.01 for pSLE patients and aSLE patients respectively with no statistical significant differences between the two studied groups (P=0.369).

*Creatinine clearance (ml/min)*

Creatinine clearance ranged from 22-89 and 20.2-90.0 with the mean of 62.1±21.4 and 52.6±24.3 for pSLE patients and aSLE patients respectively with no statistical significant differences between the two studied groups (P=0.221).

*ALT (U/L)*

ALT ranged from 10-305 and 15-322 with the mean of 45.2±50.2 and 56.9±45.8 for pSLE patients and aSLE patients respectively with no statistical significant differences between the two studied groups (P=0.098).

*AST (U/L)*

AST ranged from 10-230 and 29.5-310.0 with the mean of 43.7±39.6 and 58.0±42.1 for pSLE patients and aSLE patients respectively with no statistical significant differences between the two studied groups (P=0.211).

*ESR*

ESR ranged from 12-110 and 5-120 with the mean of 47.33±33 and 61.33±27.11 for pSLE patients and aSLE patients respectively with statistical significant differences between the two studied groups (P=0.016).

*Urinary Alb/creatinine ratio*

It ranged from 22-105 and 20-122 with the mean of 61.3±28.7 and 70.8±30.1 for pSLE patients and aSLE patients respectively with statistical significant differences between the two studied groups (P=0.041).

*CRP*

Positive CRP were found in 3 and 7, while negative CRP were found in 89 and 83 patients for pSLE patients and aSLE patients respectively, with no statistical significant differences. (P=0.0980).

aSLE patients respectively, pSLE patients have statistically higher values than aSLE patients. (P=0.021).

SLAM ranged from 10-36 and 8-27 with the mean of 18.5±6.8 and 12.6±5.98 for pSLE patients and group aSLE patients respectively, pSLE patients have statistically higher values than aSLE patients. (P=0.021).

SLEDDI ranged from 0-7 and 1-8 with the mean of 1.5±2.1 and 2.33±2.07 for pSLE patients and aSLE patients respectively, there were no statistical significant differences between the two studied groups regarding SLEDDI (P=0.074).

C3 (g/L) ranged from 0.02-1.79 and 0.11-1.98 with the mean of 0.57±0.42 and 0.71±0.51 for pSLE patients and group aSLE patients respectively, there were no statistical significant differences between the two studied groups regarding C3 (P=0.254)

C4 (g/L) ranged from 0.20-0.92 and 0.32-1.01 with the mean of 0.34±0.31 and 0.44±0.54 for pSLE patients and group aSLE patients respectively, there were no statistical significant differences between the two studied groups regarding C4 (P=0.211)

Anti-ds DNA (Iu/L) ranged from 30-396 and 25-421 with the mean of 78.6±42.6 and 112.9±51.6 for pSLE patients and aSLE patients respectively, pSLE patients have statistically higher values than aSLE patients. (P=0.038).

*Table 5. Disease activity of studied patients.*

	Group I (pSLE) "No. 92"	Group II (aSLE) "No. 90"	P
SLEDAI			
Min	4	8	0.021*
Max	60	42	
Mean	38.2	22.6	
± SD	11.2	14.2	
SLAM			
Min	10	8	0.021*
Max	36	27	
Mean	18.5	12.6	
± SD	6.8	5.98	
SLEDDI			
Min	0	1	0.074
Max	7	8	
Mean	1.5	2.33	
± SD	2.1	2.07	
C3 (g/L)			
Min	0.02	0.11	0.254
Max	1.79	1.98	
Mean	0.57	0.71	
± SD	0.42	0.51	
C4 (g/L)			
Min	0.20	0.32	0.211
Max	0.92	1.01	
Mean	0.34	0.44	
± SD	0.31	0.54	
Anti-ds DNA (Iu/L)			
Min	30	25	0.038*
Max	396	421	
Mean	78.6	112.9	
± SD	42.6	51.6	

SLEDAI = SLE disease activity index; SLAM = SLE activity measure; SLEDDI: SLE damage index.

**3.4. Disease Activity**

Table (5) shows disease activity of studied patients, it illustrated that, SLEDAI ranged from 4-60, 8-42 with the mean of 38.2±11.2 and 22.6±14.2 for pSLE patients and

**4. Discussion**

Systemic lupus erythematosus (SLE) is a complex autoimmune disease that can affect all organ systems due to alterations of both the innate and adaptive immune systems. Although onset during infancy is rare, the incidence of SLE rises steadily during childhood until mid-adulthood, especially among females. [13]

Our study focused on comparing the similarities and differences between pediatric and adult onset systemic lupus erythematosus, as well as estimating the activity indices and damage index at the time of study.

In our study, Age ranged from 8.0-16.0 and 21-52.0 years with the mean of 11.80±2.52 and 36.8±11.3 years for children and adult groups respectively and included 20 (21.7%) and 23 (25.6%) males and 72 (78.3%) and 67 (74.4%) females for pediatric and adult groups respectively.

The female to male ratio in adult onset SLE is generally found to be slightly more than 10: 1. A higher proportion of men is often reported in childhood onset SLE in some series. [14]

In our study, men represented 9% of the studied pediatric onset SLE with a female to male ratio similar to that in the adult onset SLE.

Comparison of the clinical features at onset between childhood onset and adult onset patients reveals both similarities and important differences. The frequency of skin, joint, serositis, and haematological affection were similar in both groups and correlate with previous reports. However, pediatric onset SLE patients showed increased incidence of renal involvement, fever, and lymphadenopathy, which had been reported by other authors. [15]

In the presence of suggestive clinical signs and symptoms,

in agreement with our study, laboratory testing can support and confirm the diagnosis of SLE. A hallmark of SLE is the production of multiple autoantibodies. The commonest autoantibody is the antinuclear antibody (ANA), present in more than 95% of pSLE patients. In the presence of an ANA, it is appropriate to examine for specific autoantibodies including double-stranded DNA (dsDNA) and the extractable nuclear antigens (ENAs), recognizing that particular autoantibodies correlate with certain disease features. [16] The test for ANA has high sensitivity (>95%), but its specificity for SLE is as low as 36%. [17] Moreover, up to 10% of 'healthy' children will demonstrate a positive ANA. In SLE, anti-dsDNA antibodies have high specificity. Anti-Smith antibodies (anti-Sm, not to be confused with anti-smooth muscle antibodies indicative of autoimmune hepatitis) have the greatest specificity but low sensitivity for SLE. Both anti-dsDNA and anti-Sm antibodies are associated with renal involvement, and anti-Sm may be associated with more severe disease. Other autoantibodies observed in pSLE include anti-ribonuclear protein (anti-RNP), anti-Ro (also known as anti-SSA) and anti-La (or anti-SSB) antibodies. Offspring of females with anti-Ro antibodies are at risk for Neonatal Lupus Erythematosus (NLE). NLE can lead to congenital heart block in these neonates, therefore, any adolescent female with pSLE and anti-Ro antibodies should be informed of this risk prior to any pregnancy, and referred for fetal echocardiogram monitoring by the end of the first trimester.

Other supporting features for SLE include hypocomplementemia (particularly C3 and C4 which are readily testable), cytopenia of one or more cell line as discussed earlier, and elevated ESR in the face of a normal C-reactive protein (CRP). Interestingly, CRP is often normal or only minimally elevated during a SLE flare, except when the flare is of serositis, or in the presence of concurrent infection or macrophage activation syndrome.

Our study showed that, noticeable elevations in liver enzymes, the mean of ALT were  $45.2 \pm 50.2$  and  $56.9 \pm 45.8$  and the mean of AST were  $43.7 \pm 39.6$  and  $58.0 \pm 42.1$  for pSLE and aSLE patients respectively with no statistical significant differences.

Elevated liver enzymes can indicate fatty liver (secondary to corticosteroids), an adverse drug reaction or active SLE. Less common causes in pSLE would include an intrahepatic thrombotic process, or elevated transaminases as a reflection of muscle inflammation. [17] Routine hematology and biochemistry tests are used to monitor disease status for flare and remission, medication side effects, and the effects of chronic disease and inflammation. Urine analysis should be done regularly for proteinuria, hematuria, and to examine for casts, while urine protein to creatinine ratios (spot, or 24 hour collection) are required for monitoring response to treatment of lupus nephritis. [18]

In agreement with our study, Tarr, et al., (2015) compare the clinical course of adult and pediatric-onset SLE. Data from 342 adult patients and 79 children were analyzed using hospital medical records. Organ manifestations, laboratory parameters, and immunoserological characteristics were

evaluated. They found that, gender distribution was not significantly different between both groups with disease starting in childhood vs adulthood. The prevalence of the following manifestations was significantly higher for pediatric than for adult-onset disease including: lupus nephritis (43% pediatric vs 26.4% for adult-onset), hematological disorders (57% vs 36.4%), photosensitivity (20% vs 9%), and mucosal ulceration (11.4% vs 4%). For adult-onset SLE, neurological symptoms (30% vs 6%). While this study disagreement with our study regarding polyarthritis (86% vs 68%) occurred significantly more frequently than in children. [19]

## 5. Conclusions

From our study we concluded that, both the clinical manifestations as well as serological characteristic of pediatric and adult onset SLE in Egyptian population is quite different. Pediatric onset SLE is a life long autoimmune disease that may be difficult to diagnose due to the heterogeneity of clinical presentations. It tends to lead a more active and aggressive disease course than adult onset SLE resulting in greater disease damage, increase in morbidity and mortality rates. The value of this study is to determine the long term outcome of early onset systemic lupus in addition to adopt a better tailored management, follow up and treatment approach for such young lupus patients.

---

## References

- [1] Olowu W. Childhood-onset systemic lupus erythematosus. *J Natl Med Assoc.* 2007; 99(7): 777–84.
- [2] Brunner HI, Gladman DD, Ibanez D, Urowitz MD, Silverman ED. Difference in disease features between childhood-onset and adult-onset systemic lupus erythematosus. *Arthritis Rheum.* 2008; 58(2): 556–62.
- [3] Al Salloum AA. Lupus nephritis in childhood. *Saudi J Kidney Dis Transpl.* 2003; 14(1): 43–56.
- [4] Padovan M, Govoni M, Castellino G, Rizzo N, Fotinidi M, Trotta F. Late onset systemic lupus erythematosus: no substantial differences using different cut-off ages. *Rheumatol Int.* 2007; 27(8): 735–41.
- [5] Kurahara DK, Grandinetti A, Fujii LL, Tokuda AA, Galaro JA, Han MJ, et al. Visiting consultant clinics to study prevalence rates of juvenile rheumatoid arthritis and childhood systemic lupus erythematosus across dispersed geographic areas. *J Rheumatol.* 2007; 34(2): 425–9.
- [6] Rina M, Hermine IB. Pediatric lupus-are there differences in presentation, genetics, response to therapy, damage accrual compared to adult lupus?. *Rheum Dis Clin North Am.* 2010; 36(1): 53-80.
- [7] Tucker LB, Uribe AG, Fernandez M, Vila LM, McGwin G, Apte M, et al. Adolescent onset of lupus results in more aggressive disease and worse outcomes: results of a nested matched case-control study within LUMINA, a multiethnic US cohort (LUMINA LVII) Lupus. 2008; 17(4): 314–22.

- [8] Bastian HM, Alarcon GS, Roseman JM, McGwin G, Jr, Vila LM, Fessler BJ, et al. Systemic lupus erythematosus in a multiethnic US cohort (LUMINA) XL II: factors predictive of new or worsening proteinuria. *Rheumatology (Oxford)* 2007; 46(4): 683–9.
- [9] Michelle Petri, Ana-Maria Orbai, Graciela S. Alarcón, Caroline Gordon, Joan T. Merrill, Paul R. Fortin, et al. Derivation and Validation of Systemic Lupus International Collaborating Clinics Classification Criteria for Systemic Lupus Erythematosus. *Arthritis Rheum.* 2012; 64(8): 2677–2686.
- [10] Isenberg DA, Rahman A, Allen E. Disease activity index for patients with SLE. *Rheum* 2005; 44: 902-6.
- [11] Griffiths B, Mossca M, Gordon C. Assessment of patients with systemic lupus erythematosus and the use of lupus disease activity indices. *Best pract Res Clin Rheumatol* 2005; 19(5): 685-708.
- [12] Gladman D, Grinzler E, Gold Smith C. The development and initial validation of SLE DDI ACR index for systemic lupus. *Arthritis Rheum* 1997; 40: 809-13.
- [13] Liu Z, Davidson A. Taming lupus-a new understanding of pathogenesis is leading to clinical advances. *Nat Med.* 2012; 18(6): 871-882.
- [14] Yacoub Wasef SZ. Gender differences in systemic lupus erythematosus. *Gen Med.* 2004; 1(1): 12-7.
- [15] Farber HW, Foreman AJ, Miller DP, et al. REVEAL registry: correlation of right heart catheterization and echocardiography in patients with pulmonary arterial hypertension. *Congest Heart Fail* 2011; 17: 56–64.
- [16] Jurecsek R, Fritzler M, Tyrrell P, et al. Autoantibodies in pediatric systemic lupus erythematosus: ethnic grouping, cluster analysis, and clinical correlations. *J Rheumatol.* 2009; 36: 416–21.
- [17] Deen ME, Porta G, Fiorot FJ, et al. Autoimmune hepatitis and juvenile systemic lupus erythematosus. *Lupus.* 2009; 18: 747–51.
- [18] Deborah ML, Sylvia K. Systemic lupus erythematosus in children and adolescents. *Pediatr Clin North Am.* 2012; 59(2): 345-364.
- [19] Tarr T, Dérfalvi B, Győri N, Szántó A, Siminszky Z, Malik A, Szabó AJ, Szegedi G, Zeher M. Similarities and differences between pediatric and adult patients with systemic lupus erythematosus. *Lupus.* 2015; 24(8): 796-803.