

**Comparison of Disease Characteristics, Course  
and Outcome between Adults with Juvenile and  
Adult- onset Systemic Lupus Erythematosus**

A Thesis

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# List of Abbreviations

Abb.	Meaning
ACLE.....	Acute cutaneous lupus erythematosus
ACR .....	American College of Rheumatology
AIHA .....	Autoimmune hemolytic anemia
ANA .....	Antinuclear antibody
Anti-dsDNA .....	Antinuclear double stranded DN
APL.....	Antiphospholipid antibodies
APO .....	Adverse pregnancy outcomes
APS.....	Antiphospholipid syndrome
ARDS .....	Acute respiratory distress syndrome
ASLE .....	Adult-onset SLE
AZA.....	Azathioprine
BAFF .....	B-cell activating factor
BILAG.....	British Isles Lupus Assessment Group
CHB.....	Congenital heart block
CHD .....	Coronary heart disease
CNS .....	Central nervous system
CSs.....	Corticosteroids
CVST.....	Central venous thrombosis
DHEA .....	Dehydroepiandrosterone
DM.....	Diabetes mellitus
ECG .....	Electrocardiography
ESRD.....	End stage renal disease
EULAR.....	European League Against Rheumatism
GI.....	Gastrointestinal
HAQ .....	Heath Assessment Questionnaire

# List of Abbreviations

Abb.	Meaning
IA.....	Intra-articular
IFN-alpha.....	Interferon-alpha
ILD .....	Interstitial lung disease
ITP .....	Idiopathic thrombocytopenic purpura
IUGR .....	Intra-uterine growth restriction
IVIG.....	Intravenous immunoglobulin
JSLE .....	Juvenile onset systemic lupus erythematosus
LAC .....	Lupus anticoagulant
LE .....	Lupus erythematosus
LN.....	Lupus nephritis
LVEF .....	Left ventricular ejection fraction
MTX .....	Methotrexate
MMF.....	Mycophenolate Mofetil
NPSLE .....	Neuropsychiatric lupus
NSAIDs .....	Nonsteroidal anti-inflammatory drugs
PAH .....	Pulmonary arterial hypertension
PDCs.....	Plasmacytoid dendritic cells
PML.....	Progressive multifocal leukoencephalopathy
RA.....	Rheumatoid arthritis
RNP .....	Ribonucleoprotein
SAH.....	Subarachnoid hemorrhage
SF36.....	Short Form 36
SLAM .....	Systemic Lupus Activity Measure
SLE.....	Systemic lupus erythematosus
SLEDAI.....	Systemic Lupus Erythematosus Disease Activity Index
SLICC.....	Systemic Lupus International Collaboration Clinics
TPMT .....	Thiopurine S-methyltransferase
TTP .....	Thrombocytopenic purpura

## **Abstract**

### **Background:**

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease with a broad spectrum of clinical manifestations that predominantly affects young women during childbearing age. The disease onset of SLE can also occur in pediatric and older populations.

### **Patients and Method:**

One hundred and fifty consecutive patients with SLE fulfilling the SLICC criteria for the classification of SLE (Petri et al., 2012) were included in the study. The patients were classified according to the age at disease onset.

### **Results:**

This study was conducted on One hundred and fifty patients with SLE fulfilling the SLICC criteria for the classification of SLE (Petri et al., 2012). SLE patients were 141 females (94.00%) and 9 males (6.00%).

### **Conclusion:**

- The age at onset of SLE influences its clinical and serological manifestations.
- JSLE patients have a more aggressive presentation and course than ASLE patients with a higher frequency of lupus nephritis, more cutaneous symptoms and hematological manifestations.
- JSLE patients have more severe disease activity with very high SLEDAI and SLICC damage scores.
- ASLE patients have more musculoskeletal manifestations

**Keywords:** Systemic Lupus International Collaboration, Thiopurine S-methyltransferase, Thrombocytopenic purpura

# INTRODUCTION

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease with a broad spectrum of clinical manifestations that predominantly affects young women during childbearing age (*Cervera et al., 2009*). The disease onset of SLE can also occur in pediatric and older populations (*Mak et al., 2007*).

Although the highest prevalence of SLE is among women of childbearing age, juvenile-onset SLE (JSLE) represents 10–20% of all SLE cases (*Kamphuis and Silverman E, 2010*). SLE is diagnosed early in life (before age 16) in up to 20% of cases (*Hersh et al., 2011*).

Several factors, including age and ethnicity, can affect the nature and severity of SLE, and response to treatment also varies by race, ethnicity, and age (*Borchers et al., 2010*). Childhood-onset lupus patients display some differences in their disease profile and are associated with higher disease severity, and a more-rapid damage accrual compared with adult-onset SLE (ASLE) (*Kamphuis et al., 2010*).

It has been established JSLE tends to have a more aggressive presentation and course, with high rates of organ involvement and increased need for long-term immunosuppressive medication (*Beatriz et al., 2014*). JSLE patients have more severe disease activity, and renal involvement was more frequent (*Choi et al., 2015*).

While in adults a predominance of clinical features such as serositis and sicca symptoms was observed, in adolescents

the disease was significantly more associated with the development of LN, which is considered an indicator of a worse prognosis. A notable percentage of adolescent-onset patients also developed cardiovascular events or cancer, which are uncommon diseases in young patients (*Beatriz et al., 2014*).

There are notable differences among the manifestations of the disease between children and adults. A study comparing 56 children with JSLE and 194 patients with ASLE found that renal involvement, encephalopathy and hemolytic anemia, were significantly more common in JSLE as compared to ASLE (*Hersh et al., 2009*). There is a higher prevalence of anti-dsDNA, antiSm, and anti-nucleosome antibodies in JSLE patients (*Choi et al., 2015*).

Despite a dramatic improvement in mortality in both adult and adolescent-onset disease in the last few decades, owing largely to significant advances in the therapy of SLE, patients diagnosed with SLE at an early age remain at high risk for early mortality in their young adult years (*Hersh et al., 2011*).

The poorer outcome observed in younger children may be explained by a stronger genetic predisposition, a more severe disease expression (e.g. more frequent neuropsychiatric disorders), a higher infectious susceptibility and a more aggressive therapy, particularly within the first 6 months of disease course (*Elodie et al., 2009*).