

# Clinical heterogeneity in childhood-onset systemic lupus erythematosus: Single tertiary center experience

## Çocukluk çağı başlangıçlı sistemik lupus eritematozusta klinik heterojenite: Tek üçüncü basamak merkez deneyimi

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### Abstract

**Objective:** To characterize the clinical spectrum, pattern of organ involvement, treatment strategies, and disease severity in patients with childhood-onset systemic lupus erythematosus (cSLE) at a tertiary pediatric rheumatology center.

**Methods:** This retrospective study included patients <18 years with cSLE diagnosed by Systemic Lupus International Collaborative Clinics criteria and followed between August 2019 and September 2025. Disease activity was assessed using the Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI-2K), and lupus nephritis was classified according to International Society of Nephrology/Renal Pathology Society criteria.

**Results:** The age at diagnosis was 13 years [interquartile range (IQR): 11-15]. Mucocutaneous manifestations predominated, with a cutaneous rash present in 29 patients (54.7%). Other common features included non-erosive arthritis (43.4%, n=23), oral ulcers (13.2%, n=7), and alopecia (11.3%, n=6). Renal involvement occurred in 31 patients (58.5%). Renal biopsies were conducted in 25 patients (47.1% of the study population), all of whom had histopathological confirmation of lupus nephritis. The distribution among biopsied patients was as follows: Class II: 11 (20.7%); Class IV: 11 (20.7%); Class I: 2 (3.7%); and Class V: 1 (1.8%). All patients received hydroxychloroquine. Systemic corticosteroids were administered to 50 patients (94.3%). Mycophenolate mofetil was the primary immunosuppressive treatment in 43.4% (n=23) of patients, whereas cyclophosphamide was the primary immunosuppressive treatment in 41.5% (n=22) of patients. Disease activity was high at baseline (median SLEDAI-2K score: 16, IQR: 8-20), but showed a significant improvement by the final assessment (median SLEDAI-2K score: 1.5, IQR: 0-4; p<0.001).

### Özet

**Amaç:** Üçüncü basamak bir pediatrik romatoloji merkezinde çocukluk çağı başlangıçlı sistemik lupus eritematozusta (çSLE) hastalarının klinik spektrumunu, organ tutulum paternini, tedavi stratejilerini ve hastalık şiddetini karakterize etmektir.

**Yöntem:** Bu retrospektif çalışmaya, Sistemik Lupus Uluslararası İşbirliği Klinikleri kriterlerine göre çSLE tanısı almış ve Ağustos 2019 ile Eylül 2025 arasında takip edilen 18 yaş altı hastalar dahil edildi. Hastalık aktivitesi Sistemik Lupus Eritematozusta Hastalık Aktivite İndeksi 2000 (SLEDAI-2K) ile değerlendirildi ve lupus nefriti Uluslararası Nefroloji Derneği/Böbrek Patolojisi Derneği kriterlerine göre sınıflandırıldı.

**Bulgular:** Tanı anındaki ortalama yaş 13 yıldır [çeyreklik aralık (IQR): 11-15]. Mukokutanöz bulgular baskındır ve 29 hastada (%54,7) akut veya subakut kutanöz lupus döküntüsü mevcuttur. Diğer yaygın özellikler arasında eroziv olmayan artrit (%43,4, n=23), oral ülserler (%13,2, n=7) ve alopesi (%11,3, n=6) yer aldı. Takip sırasında 31 hastada (%58,5) böbrek tutulumu meydana geldi. Yirmi beş hastaya (kohortun %47,1'i) böbrek biyopsisi yapıldı ve tüm olgularda histopatolojik olarak lupus nefriti doğrulandı. Biyopsi yapılan hastalar arasındaki dağılım şu şekildedir: 11'inde Sınıf II (%20,7), 11'inde Sınıf IV (%20,7), 2'sinde Sınıf I (%3,7) ve 1'inde Sınıf V (%1,8). Tüm hastalara (%100) hidroklorokin verildi. Elli hastaya (%91,3) sistemik kortikosteroidler uygulandı. Mikofenolat mofetil en sık kullanılan immünosüpresif ajandı (%43,4, n=23), bunu siklofosfamid (%41,5, n=22) izledi. Hastalık aktivitesi başlangıçta yüksekti (ortalama SLEDAI-2K skoru: 16, IQR: 8-20), ancak son değerlendirilmede önemli bir iyileşme gösterdi (ortalama SLEDAI-2K: 1,5, IQR: 0-4; p<0,001).

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## Abstract

**Conclusion:** This study emphasizes the heterogeneous presentation of cSLE, with frequent mucocutaneous and musculoskeletal involvement and notable renal and neurological manifestations. Early diagnosis and appropriate therapy are essential for favorable outcomes.

**Keywords:** Childhood, disease severity, lupus erythematosus, organ involvement

## Introduction

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease characterized by heterogeneous clinical presentations and broad organ involvement. Its pathogenesis involves the deposition of immune complexes and autoantibody-mediated tissue damage. Although it most commonly affects the skin and musculoskeletal system, SLE may involve any organ, including the kidneys, the central nervous system, the heart, and the lungs.<sup>[1]</sup> Approximately 15% to 20% of all SLE cases are diagnosed in pediatric patients (under 18 years of age), a subgroup formally termed childhood-onset SLE (cSLE). Although the disease usually appears after the first decade of life, it can rarely begin before age 5; findings in this early period require consideration of monogenic forms of SLE.<sup>[2,3]</sup>

Comparative analyses of adult-onset SLE and cSLE have revealed that different clinical profiles may exist. cSLE is characterized by a significantly higher prevalence of constitutional and mucocutaneous manifestations—including fever, lymphadenopathy, and malar rash—as well as hematological abnormalities, such as cytopenias.<sup>[4-6]</sup> Furthermore, renal involvement, particularly lupus nephritis, is reported more frequently in cSLE; however, the distribution of histopathological subtypes appears similar to that observed in adults.<sup>[7,8]</sup> Neuropsychiatric manifestations of SLE (NPSLE) have been reported more commonly in cSLE, although their exact prevalence remains uncertain.<sup>[6,9]</sup>

The diagnosis of cSLE is established using validated classification systems, including those from the American College of Rheumatology (ACR), the Systemic Lupus International Collaborating Clinics (SLICC), and the European Alliance of Associations for Rheumatology/ACR 2019 criteria.<sup>[10-12]</sup> Symptoms and age at diagnosis can influence the disease phenotype, organ involvement, and serological characteristics.<sup>[13]</sup> Advances in diagnostic techniques and increased disease awareness have contributed to improved outcomes compared with earlier reports.<sup>[14]</sup> Nevertheless, cSLE tends to present with higher disease activity and severity, primarily due to the increased frequency of major organ involvement, such as lupus nephritis.<sup>[15]</sup> The management of cSLE requires a multidisciplinary approach aimed at controlling disease activity, preventing flares and

## Özet

**Sonuç:** Bu çalışma, sık görülen mukokutanöz ve kas-iskelet sistemi tutulumu ve belirgin renal ve nörolojik bulgularla birlikte cSLE'nin heterojen bir tablo sergilediğini vurgulamaktadır. Olumlu sonuçlar için erken tanı ve uygun tedavi esastır.

**Anahtar Kelimeler:** Çocukluk çağı, hastalık şiddeti, lupus eritematozus, organ tutulumu

irreversible organ damage, achieving remission, and minimizing treatment-related adverse effects.<sup>[16]</sup>

The aim of this study is to present the clinical features, disease activity and severity, and disease management of cSLE patients followed at a tertiary pediatric rheumatology center.

## Materials and Methods

This retrospective longitudinal study was conducted at our institution between August 2019 and September 2025 in patients aged <18 years who met SLICC criteria and were followed with a diagnosis of cSLE. Patients with missing data or a follow-up period shorter than 6 months were excluded from the study.

Data were systematically collected on demographic characteristics, clinical findings, and organ system involvement. Recorded laboratory data included hematological parameters (hemoglobin levels, leukocyte counts, and platelet counts) and inflammatory indicators, specifically C-reactive protein and erythrocyte sedimentation rate. The immunological assessment focused on the presence of antinuclear antibodies (ANA), anti-double-stranded DNA (anti-dsDNA), and extractable nuclear antigens (ENA). Additionally, pathological data, particularly renal biopsy results for patients with lupus nephritis, were documented. Disease activity was quantified at diagnosis and at the most recent clinical evaluation using the Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI-2K).<sup>[17,18]</sup> The histopathological classification of lupus nephritis was conducted in accordance with the International Society of Nephrology/Renal Pathology Society (ISN/RPS) recommendations.<sup>[19]</sup>

Antiphospholipid antibody (aPL) positivity was defined as the persistent presence of lupus anticoagulant, anticardiolipin [immunoglobulin G (IgG)/immunoglobulin M (IgM)], and/or anti- $\beta$ 2 glycoprotein I (IgG/IgM) antibodies on at least two occasions, measured  $\geq$ 12 weeks apart. Pediatric antiphospholipid syndrome (APS) was defined according to the revised 2006 Sapporo criteria, which require at least one clinical event (documented arterial, venous, or small-vessel thrombosis) and at least one persistently positive aPL test on two occasions at least 12 weeks apart.<sup>[20]</sup>

For the assessment of SLE disease activity, remission was categorized as off-treatment or on-treatment. Off-treatment

remission was defined as an SLEDAI-2K score of 0 in the absence of systemic corticosteroids and immunosuppressive therapy, whereas on-treatment remission was defined as an SLEDAI-2K score of 0 with prednisolone  $\leq 5$  mg/day and maintenance-dose immunosuppressive therapy. Low disease activity state was defined as an SLEDAI-2K score  $< 4$  with prednisolone  $< 7.5$  mg/day and maintenance-dose immunosuppressive therapy, whereas high disease activity state was defined as an SLEDAI-2K score  $> 4$  with prednisolone  $> 7.5$  mg/day and induction-dose immunosuppressive therapy.<sup>[21]</sup>

At our center, renal biopsy is performed in pediatric SLE patients in the presence of significant proteinuria (including nephrotic-range proteinuria) persistent hematuria, reduced renal function, or rapidly progressive glomerulonephritis. Biopsy is also considered in cases of persistent active lupus nephritis despite therapy or when clinical findings suggest subclinical renal involvement.

The study was approved by University of Health Sciences Türkiye, Ankara Bilkent City Hospital's Ethical Committee (approval number: TABED1-25-1785, date: 22.10.2025) and conducted in accordance with the Declaration of Helsinki.

### Statistical Analysis

Statistical analyses were conducted using SPSS software, version 20.0 (IBM Corp., Armonk, NY, USA). Descriptive statistics were reported as counts (n) for categorical data and as means, medians, and standard deviations for continuous variables. The Shapiro-Wilk test was employed to assess the normality of data distribution. For group comparisons, independent Student's t-tests were used for normally distributed variables, and Mann-Whitney U tests were used for non-normally distributed variables. Statistical significance was defined as  $p < 0.05$ .

### Results

A total of 53 patients diagnosed with cSLE according to the SLICC classification criteria were enrolled in this study. The cohort was predominantly female (n=48, 90.6%), corresponding to a female-to-male ratio of approximately 9:1. The median age at cSLE diagnosis was 13 years (range: 11-15 years), and the median follow-up duration was 26 months (range: 12-48 months) (Table 1).

Mucocutaneous involvement was a common feature, with an acute or subacute cutaneous lupus rash observed in 29 patients (54.7%). Other frequent manifestations included non-erosive arthritis (n=23, 43.4%), oral ulcers (n=7, 13.2%), and alopecia (n=6, 11.3%). All clinical findings and their frequencies are given in Table 1.

Serological profiling revealed a high prevalence of autoantibodies. ANA was detected in 51 (96.2%) of patients,

and anti-dsDNA was positive in 43 (81.1%) of patients. Hypocomplementemia, defined by low C3 and/or C4 levels, was observed in 39 (73.6%) of the study population. Antiphospholipid antibodies were present in 11 patients (21.6%), and 8 patients (15.1%) met the criteria for aPL syndrome. Raynaud's phenomenon was noted in two patients (3.7%). A summary of immunological characteristics is presented in Table 1.

Hematological abnormalities were common, with anemia detected in 18 patients (33.9%), lymphopenia in 16 (30.1%), and thrombocytopenia in 5 (9.4%). Pancytopenia was documented in four patients (7.5%).

Renal involvement developed in 31 patients (58.5%) during the follow-up period. A renal biopsy was performed in 25 patients (47.1% of the total cohort); all biopsies confirmed lupus nephritis. Histopathological classification, according to the ISN/RPS 2003 criteria, revealed the following distribution among biopsied cases: Class II in 11 cases (20.7%), Class IV in 11 cases (20.7%), Class I in 2 cases (3.7%), and Class V in 1 case (1.8%).

Neurological involvement was observed in 19 patients (33.9%), with mood changes being the most common manifestation (n=10, 18.2%), followed by seizures (n=3, 5.6%), psychosis (n=2, 3.7%), neuropathy (n=2, 3.7%), mononeuritis multiplex (n=1, 1.8%), and myelitis (n=1, 1.8%).

The cohort exhibited high disease activity at baseline, with a median SLEDAI-2K score of 16 (IQR: 8-20). At the final assessment, a significant reduction was observed, with the median SLEDAI-2K score decreasing to 1.5 (IQR 0-4;  $p < 0.001$ ), as detailed in Table 1.

Hydroxychloroquine was universally administered (n=53, 100%). Systemic corticosteroids were used in 50 patients (94.3%). Regarding immunosuppressive agents, mycophenolate mofetil (MMF) was the most frequently prescribed (n=23, 43.4%), followed by cyclophosphamide (n=22, 41.5%). In a subset of eight patients with renal involvement, rituximab was administered following cyclophosphamide treatment. Plasmapheresis was performed in the management of 4 patients (7.5%) (Table 1).

Comorbid conditions, distinct from manifestations attributable to cSLE, were present in 30 patients (56.6%) at the time of diagnosis. The spectrum of documented comorbidities included epilepsy (n=4), delayed puberty or amenorrhea (n=4), hepatitis or cholestasis (n=3), Hashimoto's thyroiditis (n=2), type 1 diabetes mellitus (n=1), and Klippel-Feil syndrome (n=1) (Table 2).

### Discussion

This study delineates the clinical, serological, and histopathological profiles of cSLE within a single tertiary pediatric rheumatology referral center. While mucocutaneous and musculoskeletal symptoms were the most prevalent clinical manifestations, renal and neurological complications

were the primary organ systems involved. Within the lupus nephritis subgroup, class II and IV emerged as the predominant histopathological patterns. Furthermore, the substantial decline in initially high SLEDAI-2K scores throughout the follow-up period reflects the efficacy of the administered immunosuppressive regimens in achieving disease stabilization.

cSLE is a multisystem inflammatory disorder marked by pronounced heterogeneity in its clinical presentation. The disease spans a broad clinical spectrum, ranging from relatively mild manifestations to severe, life-threatening organ

involvement. Many initial symptoms, including oral ulcers, fever, arthralgia, headache, and weight loss, are non-specific and commonly overlap with other childhood illnesses, complicating early diagnosis. Notably, the incidence of major organ involvement—such as arthritis, nephritis, and neuropsychiatric manifestations—is inversely related to age at disease onset.<sup>[22,23]</sup> A Turkish cohort study demonstrated that patients with cSLE exhibit an increased prevalence of involvement of the renal, mucocutaneous, hematologic, and neuropsychiatric organ systems, coupled with elevated seropositivity rates for anti-dsDNA and anticardiolipin antibodies.<sup>[24]</sup> These observations are supported by parallel findings from a Canadian cohort, which reported significantly elevated frequencies of neuropsychiatric and anticardiolipin antibody positivity in patients with cSLE.<sup>[25]</sup> Furthermore, cross-national epidemiological studies conducted in France and China consistently report that pediatric-onset disease is associated with higher rates of renal and hematologic involvement, as well as overall greater disease severity compared with adult-onset SLE. Collectively, these distinctions not only highlight the critical need for early recognition and intervention in cSLE, but also point to fundamental challenges in applying classification criteria developed primarily for adult populations to pediatric cases.<sup>[26,27]</sup>

Renal involvement is the most prevalent and serious organ manifestation in cSLE, occurring more frequently than in adult-onset disease. Epidemiological studies report prevalence estimates for lupus nephritis in cSLE ranging from 30% to 75%.<sup>[28]</sup> A definitive diagnosis requires histopathological confirmation via renal biopsy, a procedure recommended when renal disease is clinically suspected in SLE patients.<sup>[29]</sup> While immune complex-mediated glomerulonephritis is the most common lesion, the

**Table 1. Demographic, clinical, laboratory and disease severity characteristics of patients with childhood lupus erythematosus (n=53)**

Variables	
Gender, female (n, %)	48 (90.6%)
Age at diagnosis, (years, min-max)	13 (11-15)
Follow-up period, (months, min-max)	26 (12-48)
Clinical findings (n, %)	
Constitutional symptoms	37 (69.8%)
Cutaneous involvement	29 (54.7%)
Arthritis	23 (43.4%)
Oral ulcer	7 (13.2%)
Alopecia	6 (11.3%)
Serositis	19 (35.8%)
Renal involvement	31 (58.5%)
Neurological involvement	19 (33.9%)
Gastrointestinal involvement	6 (11.3%)
Cardiac involvement	8 (15.1%)
Lung involvement	10 (18.8%)
Antiphospholipid antibody syndrome	8 (15.1%)
Raynaud phenomenon	2 (3.7%)
Laboratory findings (n, %)	
Hypocomplementemia	39 (73.6%)
Direct Coombs positivity	33 (68.8%)
ANA positivity	51 (96.2%)
Anti-dsDNA positivity	43 (81.1%)
Antiphospholipid antibody positivity	11 (21.6%)
Remission status (n, %)	38 (71.7%)
Treatment (n, %)	
Hydroxychloroquine	53 (100%)
Corticosteroids	50 (94.3%)
Mycophenolate mofetil	23 (43.4%)
Cyclophosphamide	22 (41.5%)
Intravenous immunoglobulin	14 (26.4%)
Rituximab	8 (15.1%)
Plasmapheresis	4 (7.5%)
SLEDAI-2K score at diagnosis (median, min-max)	16 (8-20)
Last visit SLEDAI-2K score (median, min-max)	1.5 (0-4)
ANA: Antinuclear antibodies, min-max: Minimum-maximum, SLEDAI-2K: Systemic Lupus Erythematosus Disease Activity Index 2000	

**Table 2. Comorbidities in childhood lupus erythematosus patients in our cohort**

Comorbid diseases	n (%)
Epilepsy	4 (7.5%)
Delayed puberty, amenorrhea	4 (7.5%)
Hepatitis, cholestasis	3 (5.4%)
Hashimoto's thyroiditis	2 (3.7%)
Short stature	2 (3.7%)
Insulin resistance	2 (3.7%)
Obesity	2 (3.7%)
Glial tumor	2 (3.7%)
Precocious puberty	1 (1.8%)
Type 1 diabetes mellitus	1 (1.8%)
Secondary Cushing's syndrome	1 (1.8%)
Psoriasis	1 (1.8%)
Migraine	1 (1.8%)
Asthma	1 (1.8%)
Klippel Feil syndrome	1 (1.8%)

histopathological spectrum also includes tubulointerstitial nephritis, lupus podocytopathy, and various forms of renal vascular injury.<sup>[30]</sup> Akgun et al.<sup>[31]</sup> found the incidence of lupus nephritis to be 43.2%. Class IV lupus nephritis, the most common subtype, was the most frequently observed class in our study. In one study, renal involvement was observed in 115 (53.2%) of 216 patients in a cSLE cohort. Lupus nephritis developed in 85 of these patients. Among those who developed nephritis, class IV was the most common.<sup>[24]</sup> Renal involvement was detected in 31 patients (58.5%) of our cohort. Of these patients, eleven had class II and the remainder had class IV lupus nephritis.

Neuropsychiatric involvement is more frequent and is associated with substantial morbidity in cSLE. The clinical spectrum is highly diverse, encompassing manifestations ranging from headaches and cognitive dysfunction to psychosis and cerebrovascular events. These neuropsychiatric features can serve as the initial presentation of the disease and are often correlated with poorer long-term outcomes. The diagnosis and management of NPSLE remain particularly challenging in the pediatric population.<sup>[22]</sup> Artim-Esen et al.<sup>[24]</sup> reported a higher prevalence of neuropsychiatric involvement in the cSLE group (12.1%) and less associated damage than in the adult-onset SLE group. Neuropsychiatric involvement was observed in 19 (35.8%) of the 53 patients in the cohort. The most common diagnosis in this group was mood swings, observed in 10 patients.

Hematologic abnormalities are a common feature of cSLE, with autoimmune cytopenias representing a frequent finding. The spectrum of involvement includes leukopenia, lymphopenia, neutropenia, thrombocytopenia, and anemia. In clinical practice, during the diagnostic evaluation, it is essential to exclude other potential causes of cytopenias, such as nutritional deficiencies, concurrent infections, or medication effects.<sup>[22]</sup> In two studies published in Türkiye, Artim-Esen et al.<sup>[24]</sup> reported that the most common hematological abnormality in their cohort was lymphopenia (59.7%), followed by anemia (33.3%), and thrombocytopenia (29.6%). In contrast, Akgun et al.<sup>[31]</sup>, similar to our study, reported that the most common abnormalities were anemia (51.6%), lymphopenia (37.8%), and thrombocytopenia (21.6%), respectively. All patients in the cohort had hematologic involvement. Anemia was the most common (n=18, 33.9%), followed by lymphopenia (n=16, 30.1%) and thrombocytopenia (n=15, 28.3%). Pancytopenia was observed in 4 patients (14.3%).

The presence of antiphospholipid antibodies is associated with hypercoagulability, predisposing patients to thrombosis and thromboembolism. Additionally, systemic manifestations, such as lymphadenopathy and hepatosplenomegaly, may be observed; in severe cases, macrophage activation syndrome, a life-threatening complication can develop.<sup>[32]</sup> In a previous study of patients tested for antiphospholipid antibodies, positivity for

aCL (IgM and/or IgG) was observed in 12% of patients. However, only three of these seropositive patients (3.3% of the total cohort) met the diagnostic criteria for aPL syndrome.<sup>[5]</sup> While aPL positivity was observed in 11 patients in the cohort, APS was detected in 8 of these patients.

Mucocutaneous involvement is a common clinical feature of cSLE. Cardinal manifestations include photosensitive malar rash, discoid lesions, non-scarring alopecia, and painless oral or nasal ulcers. The recognition of these cutaneous signs is diagnostically crucial, as identifying lupus-specific rashes can facilitate earlier diagnosis and help prevent delays in disease management.<sup>[22]</sup> In a cohort of 512 patients with cSLE, Zhang et al.<sup>[33]</sup> identified cutaneous lupus as the most common mucocutaneous manifestation (29.3%), followed by oral ulcers (23.1%) and non-scarring alopecia (19.5%). Cutaneous involvement was observed in 29 patients (54.7%). Oral ulcers were observed in 7 (13.2%) patients, and regional alopecia in 6 (11.3%).

The management of cSLE poses considerable challenges, largely attributable to the marked clinical heterogeneity among patients. Although treatment guidelines from various rheumatology societies are primarily derived from adult studies, they are routinely applied to pediatric cases, including guidelines for lupus nephritis.<sup>[34-36]</sup> A cornerstone of these recommendations is the universal administration of hydroxychloroquine, unless contraindications exist.<sup>[37]</sup> Treatment intensity is typically escalated based on organ involvement through the use of corticosteroids, conventional disease-modifying antirheumatic drugs (DMARDs), and other immunosuppressive agents. This finding aligns with a UK cohort study that identified MMF as the predominant immunosuppressive agent.<sup>[38]</sup> A recent study reported that all but one patient used hydroxychloroquine, and the most commonly used DMARD was MMF, accounting for 51.3% of cases.<sup>[31]</sup> In our study, MMF was the most frequently used DMARD, accounting for 43.3% of cases. For lupus nephritis, specific induction therapy for proliferative disease involves cyclophosphamide or MMF in combination with corticosteroids. Recent guideline updates indicate that belimumab or calcineurin inhibitors added to standard of care may also be considered first-line options, though patient selection criteria remain undefined.<sup>[36]</sup> Consequently, in the absence of definitive treatment algorithms, institutional experience and center-specific protocols play a decisive role in therapeutic decision-making.

Adherence to established treatment guidelines and the implementation of early, aggressive therapeutic strategies are associated with significant improvements in disease activity metrics. This is evidenced by longitudinal studies; for instance, one Turkish cohort reported a decline in SLEDAI-2K scores from a mean of 22.5±8.1 at diagnosis to a final median of 0. Similarly, a 2021 multinational study of 670 cSLE patients documented a reduction in the mean SLEDAI-2K score from 16.5±8.9 at baseline to 4.6±5.8

at the final assessment.<sup>[39,40]</sup> The SLEDAI-2K score for patients in the cohort was 16 (8-20) at diagnosis and 1.5 (0-4) at last follow-up.

### Study Limitations

Certain limitations of the current study warrant consideration. Primarily, its retrospective nature and the relatively modest sample size may limit the generalizability of the conclusions. As this was a single-center investigation focused on a rare pediatric condition, the distribution of lupus nephritis classes may not fully align with the broader literature, which could affect the generalizability of our results. Conversely, a major strength of this research is the comprehensive documentation of disease activity trajectories and long-term patient outcomes, which are critical for managing a high-morbidity disease such as cSLE.

### Conclusion

This study highlights the heterogeneous clinical spectrum of cSLE, characterized by frequent mucocutaneous and musculoskeletal involvement and by significant renal and neurological involvement. Despite high initial disease activity, favorable outcomes were achieved through timely diagnosis and appropriate immunosuppressive therapy. These findings underscore the importance of early diagnosis, close follow-up, and multidisciplinary management in optimizing long-term outcomes in cSLE.

### Ethics

**Ethics Committee Approval:** The study was approved by University of Health Sciences Türkiye, Ankara Bilkent City Hospital's Ethical Committee (approval number: TABED1-25-1785, date: 22.10.2025) and conducted in accordance with the Declaration of Helsinki.

**Informed Consent:** Retrospective study.

### Footnotes

#### Authorship Contributions

Surgical and Medical Practices: Ş.E., U.S.B., B.Ç.A., Concept: Ş.E., U.S.B., B.Ç.A., Design: Ş.E., U.S.B., B.Ç.A., Data Collection and Processing: Ş.E., E.Ö., Ş.E.T., D.Ö., M.I.E., Y.U.E., S.N.Y., N.Ç.P., E.E., Analysis or Interpretation: Ş.E., Z.E.T., Literature Search: Ş.E., N.Ç.P., U.S.B., B.Ç.A., Writing: Ş.E.

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