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Original Research Article

CLINICAL MANIFESTATIONS AND DISEASE ACTIVITY IN CHILDHOOD SYSTEMIC LUPUS ERYTHEMATOSUS AT A TERTIARY CARE CENTRE

Ranjitha M.[1], Anusha S[2], Anil Kumar Tennelli[3], Alkarani T. Patil[4], Jacqueline A. Shah[5]

[1]Assistant Professor, Department of Pediatric Medicine, Indira Gandhi Institute of Child Health, Bengaluru, Karnataka, India.
[2]Consultant Paediatrician, ESI Hospital, Davangere, Karnataka, India.
[3]Assistant Professor, Department of Paediatrics, Indira Gandhi Institute of Child Health, Bengaluru, Karnataka, India.
[4]Professor & Head of Department, Department of Paediatrics, Indira Gandhi Institute of Child Health, Bengaluru, Karnataka, India.
[5]Assistant Professor, Department of Paediatrics, Indira Gandhi Institute of Child Health, Bengaluru, Karnataka, India.

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Corresponding Author:
Dr. Anil Kumar Tennelli
Assistant Professor, Department of Paediatrics, Indira Gandhi Institute of Child Health, Bengaluru, Karnataka, India.
Email: tennellikumar@gmail.com

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ABSTRACT

Background: Childhood-onset systemic lupus erythematosus is a multisystem autoimmune disorder that may present with high disease activity and major-organ involvement. This study assessed the clinical manifestations and disease activity of children with SLE at diagnosis and during follow-up.

Materials and Methods: This prospective observational study included 26 children aged 1–18 years fulfilling SLICC criteria for SLE at a tertiary care centre. Clinical manifestations, hematological and immunological parameters, renal involvement, and histopathological findings were recorded. Disease activity was assessed using SLEDAI-2K at diagnosis and six months.

Results: The mean age at presentation was approximately 12 years, and 84.6% were females. Serositis (65.4%) was the most frequent clinical manifestation, while hematological abnormalities were prominent. Renal and neurological involvement occurred in 42.3% and 46.2%, respectively. ANA was positive in 84.6% and anti-dsDNA in 65.4%. Among biopsy-proven lupus nephritis cases, Class IV was most common (66.7%). At diagnosis, 80.8% had high disease activity. Among survivors, mean SLEDAI-2K decreased from 24.73 ± 11.46 at diagnosis to 2.92 ± 3.52 at six months. At six months, 13 (50.0%) children were alive, 11 (42.3%) had died, and two (7.7%) were lost to follow-up.

Conclusion: Childhood SLE was characterized by female predominance, substantial multisystem involvement, and high disease activity at presentation. Disease activity improved considerably among survivors; however, the high early mortality highlights the need for timely diagnosis, intensive treatment of severe disease, and close follow-up.

Keywords: Childhood systemic lupus erythematosus; cSLE; SLEDAI-2K; lupus nephritis; disease activity; clinical manifestations.

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disorder characterized by loss of immune tolerance, autoantibody production, and multisystem inflammation. Childhood-onset SLE (cSLE), generally defined as SLE presenting before 18 years of age, accounts for a smaller proportion of overall SLE but often follows a more aggressive course than adult-onset disease, with substantial heterogeneity across geographic and ethnic populations.[1] Children may present with constitutional, mucocutaneous, musculoskeletal, hematological, renal, neurological, and cardiopulmonary manifestations, making early recognition challenging.[2]

Compared with adult disease, cSLE is frequently associated with greater disease activity and a higher burden of major organ involvement, particularly lupus nephritis, hematological abnormalities, and neuropsychiatric manifestations.[3] Early identification of these manifestations is important because childhood SLE may result in considerable morbidity, treatment-related complications,

cumulative organ damage, and mortality.[4] Contemporary international efforts therefore emphasize systematic characterization and standardized assessment of clinical and laboratory features in children with SLE.[5]

Disease severity at presentation and its subsequent course vary considerably among affected children.[6] Objective assessment of disease activity is consequently important for therapeutic decisions and follow-up. The Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI-2K) provides a standardized assessment of disease activity, while current pediatric treat-to-target approaches emphasize achieving sustained low disease activity or remission to improve long-term outcomes.[7] Hence, the present study was undertaken to assess the clinical manifestations and disease activity at diagnosis and follow-up among children with SLE at a tertiary care centre.

MATERIALS AND METHODS

Study Design and Setting

This was a **prospective observational study** conducted at the **Indira Gandhi Institute of Child Health, Bengaluru**, a tertiary care pediatric centre, over a period of **18 months**.

Study Population

Children aged **1–18 years** who were newly diagnosed with systemic lupus erythematosus and fulfilled the **Systemic Lupus International Collaborating Clinics (SLICC) classification criteria** were enrolled in the study. Written informed consent was obtained from the parents/guardians of children up to 7 years of age, while assent was obtained from children aged 8–18 years as applicable.

Inclusion Criteria

All children below 18 years of age with a new diagnosis of SLE fulfilling the SLICC classification criteria and for whom informed consent/assent was available were included.

Exclusion Criteria

Children who were previously diagnosed cases of SLE and attending follow-up, those with **drug-induced lupus**, and those with **mixed connective tissue disorder** were excluded.

Sample Size

The sample size was estimated using an anticipated prevalence of childhood SLE of **20%**, a 95% confidence level (Z=1.96), and an absolute precision of **18%**. The minimum calculated sample size was **20 children**. During the study period, **26 eligible children** were ultimately included in the analysis. The assumptions and calculated minimum sample size are documented in the study protocol.

Data Collection and Clinical Assessment

For each participant, demographic and clinical information including **age, sex, anthropometric measurements, and duration of symptoms** was recorded. Detailed clinical examination evaluated fever, malar and discoid rash, photosensitivity, alopecia, oral ulceration, vasculitis, nephritis, myositis, serositis, gastrointestinal and pulmonary manifestations, and neurological involvement including headache, seizures, psychosis, visual disturbances, and cranial nerve abnormalities.

Laboratory Evaluation

Laboratory investigations included hemoglobin, total and differential leukocyte counts, platelet count, erythrocyte sedimentation rate, C-reactive protein, renal and liver function tests, antinuclear antibody (ANA), anti-double-stranded DNA (anti-dsDNA) antibodies, complement C3 levels, urine routine examination, and urinary protein-to-creatinine ratio.

Renal biopsy was performed in children with evidence of renal involvement, including significant proteinuria or elevated serum creatinine. Histopathological findings were classified according to the **International Society of Nephrology classification for lupus nephritis**.

Assessment of Disease Activity and Follow-up

Disease activity was assessed at diagnosis using the **SLEDAI-2K scoring system**, which evaluates 24 clinical and laboratory features across neurological, vascular, renal, musculoskeletal, serosal, dermal, immunological, constitutional, and hematological domains.

Disease activity was categorized as **remission (score 0), low disease activity (0–5), moderate disease activity (6–12), and high disease activity (>12)** according to the study protocol. Children received appropriate immunosuppressive therapy based on disease type and severity and were followed for **6 months**, when clinical outcome and SLEDAI-2K disease activity were reassessed.

Statistical Analysis

Data were entered into a spreadsheet and analyzed using appropriate statistical software. Continuous variables were summarized as **mean ± standard deviation (SD)** or median with interquartile range, as appropriate, while categorical variables were expressed as **frequency and percentage**. Associations between categorical variables were assessed using the **Chi-square test or Fisher's exact test** where appropriate. Changes in disease activity between diagnosis and six-month follow-up were assessed using appropriate paired statistical tests. A **p-value <0.05** was considered statistically significant.

RESULTS

A total of 26 children with systemic lupus erythematosus (SLE) were enrolled and analyzed. The age at presentation ranged from 5 to 17 years, with a mean age of approximately 12 years. Most children (69.2%) were aged ≥10 years. There was a marked female predominance, with 22 females (84.6%) and 4 males (15.4%), giving a female-to-male ratio of approximately 5.5:1. The mean age at

onset was 11.80 years, while the mean age at diagnosis was 12.11 years.

Table 1: Demographic and baseline characteristics of children with SLE (N=26)

Characteristic	Value
Age at presentation, range (years)	5–17
Mean age at presentation (years)	12.0
Age <10 years, n (%)	8 (30.8)
Age ≥10 years, n (%)	18 (69.2)
Female, n (%)	22 (84.6)
Male, n (%)	4 (15.4)
Female:male ratio	5.5:1
Mean age at onset (years)	11.80
Mean age at diagnosis (years)	12.11
Mean weight (kg)	29.20
Mean height (cm)	138.07
Mean BMI (kg/m²)	14.55

Among the clinical manifestations, serositis was the most frequent, occurring in 17 (65.4%) children, followed by neurological involvement in 12 (46.2%), renal involvement in 11 (42.3%), synovitis in 10 (38.5%), and acute cutaneous lupus in 9 (34.6%). Hematological involvement was prominent, with thrombocytopenia reported in 19 (73.1%), leukopenia in 15 (57.7%), and hemolytic anemia in 6 (23.1%) children.

Table 2: Clinical and immunological manifestations among children with SLE

Manifestation/parameter	n	%
Clinical manifestations		
Serositis	17	65.4
Neurological involvement	12	46.2
Renal involvement	11	42.3
Synovitis	10	38.5
Acute cutaneous lupus	9	34.6
Non-scarring alopecia	7	26.9
Oral ulcers	12	46.2
Chronic cutaneous lupus	1	3.8
Hemolytic anemia	6	23.1
Leukopenia	15	57.7
Thrombocytopenia	19	73.1
Immunological findings		
ANA positive	22	84.6
Anti-dsDNA positive	17	65.4
Anti-Sm positive	5	19.2
APLA positive	3	11.5
Low C3	16	61.5
Direct Coombs test positive	9	34.6
Biopsy-proven lupus nephritis	9	34.6

ANA positivity was the most frequent immunological finding (84.6%), followed by anti-dsDNA positivity (65.4%) and low C3 levels (61.5%). Nine children (34.6%) had biopsy-proven lupus nephritis. Among these, Class IV lupus nephritis was the predominant histological pattern (66.7%), followed by Class V (22.2%) and Class III (11.1%).

Table 3: Disease activity according to SLEDAI-2K at diagnosis and six-month follow-up

SLEDAI-2K category	At diagnosis, n (%)	At 6 months, n (%)
High disease activity (>12)	21 (80.8)	1 (3.8)
Moderate disease activity (6–12)	4 (15.4)	3 (11.5)
Low disease activity (0–5)	1 (3.8)	3 (11.5)
Remission	0	6 (23.1)
Death	—	11 (42.3)
Lost to follow-up	—	2 (7.7)
Total	**26 (100)**	**26 (100)**

At diagnosis, the majority of children had high disease activity (80.8%), while 15.4% had moderate and only 3.8% had low disease activity. Among the 13 surviving children evaluated at six months, disease activity decreased substantially: six achieved remission, three had low disease activity, three had moderate disease activity, and only one continued to have high disease activity. The mean SLEDAI-2K score decreased from 24.73 ± 11.46 at diagnosis to 2.92 ± 3.52 at six months among survivors.

Table 4: Six-month outcome and its relationship with disease activity

Outcome parameter	Result
Alive at 6 months, n (%)	13 (50.0)
Death, n (%)	11 (42.3)
Lost to follow-up, n (%)	2 (7.7)
Remission among survivors, n/N (%)	6/13 (46.2)
Mean SLEDAI-2K at onset – survivors	22.54 ± 12.31
Mean SLEDAI-2K at onset – deaths	24.82 ± 10.01
Mean SLEDAI-2K at onset – lost to follow-up	38.50 ± 4.95
High SLEDAI at onset among survivors, n/N (%)	10/13 (76.9)
High SLEDAI at onset among deaths, n/N (%)	9/11 (81.8)
Association of baseline SLEDAI category with 6-month outcome	χ^2=1.48; p=0.830

At six months, 13 (50.0%) children were alive, 11 (42.3%) had died, and 2 (7.7%) were lost to follow-up. Nine of the 11 deaths (81.8%) occurred among children who had high disease activity at presentation; however, the association between baseline SLEDAI-2K category and six-month outcome was not statistically significant (p=0.830).

Overall, the findings demonstrate that cSLE presented predominantly in adolescent females, with frequent serosal, hematological, neurological, and renal involvement. Most children had high disease activity at diagnosis. Although a marked reduction in SLEDAI-2K scores was observed among survivors following treatment, the cohort experienced substantial early mortality during the six-month follow-up period.

DISCUSSION

Childhood-onset systemic lupus erythematosus (cSLE) is generally characterized by greater disease activity and more frequent major-organ involvement than adult-onset disease. In the present study, 26 children with SLE were evaluated, with a mean age at presentation of approximately 12 years and a marked female predominance (84.6%). Most patients (80.8%) had high disease activity at diagnosis, emphasizing the severe presentation of cSLE in a tertiary-care setting. Disease activity assessment is particularly important because achieving low disease activity has emerged as an important therapeutic target in childhood SLE.[8]

Serositis was the most frequent clinical manifestation (65.4%), followed by neurological (46.2%) and renal involvement (42.3%). Hematological abnormalities were also prominent, particularly thrombocytopenia and leukopenia. The multisystem pattern observed in our cohort reinforces the heterogeneous nature of cSLE. Gotch et al. demonstrated that active implementation of a low disease activity state as a therapeutic endpoint in routine pediatric SLE practice was feasible and associated with better clinical outcomes.[9]

ANA was positive in 84.6% of patients, anti-dsDNA antibodies in 65.4%, and low C3 levels were frequently observed. These findings indicate considerable immunological activity at presentation. Wahadat et al. similarly demonstrated that lupus low disease activity state is an attainable treatment target in cSLE and emphasized the importance of controlling both clinical and serological disease activity.[10] The substantial disease burden in the present cohort also supports the need for early recognition and appropriately intensive therapy, as highlighted in contemporary reviews of juvenile-onset SLE management.[11]

Renal involvement occurred in 42.3% of children, while nine patients had biopsy-proven lupus nephritis. Among biopsied patients, Class IV lupus nephritis was the predominant histological pattern (66.7%), followed by Class V (22.2%) and Class III (11.1%). The high frequency of proliferative nephritis is clinically important because renal disease represents a major determinant of long-term morbidity in cSLE. The SHARE recommendations emphasize prompt recognition, renal biopsy where indicated, and appropriate immunosuppressive treatment for childhood lupus nephritis.[12]

The diagnosis in the present study was based on SLICC criteria, which incorporate a broad spectrum of clinical and immunological manifestations and allow classification in the presence of biopsy-proven lupus nephritis with appropriate serological evidence.[13] Disease activity was assessed using SLEDAI-2K, a standardized instrument suitable for longitudinal evaluation of SLE activity.[14] Among survivors, the mean SLEDAI-2K score declined markedly from 24.73 ± 11.46 at diagnosis to 2.92 ± 3.52 at six months, indicating substantial improvement following treatment.

Despite improvement among survivors, mortality was considerable, with 11 (42.3%) children dying within six months. Nine of these 11 deaths had high-grade disease at presentation, although baseline SLEDAI category was not significantly associated with six-month outcome (p=0.830), possibly because of the small sample size. Current EULAR recommendations emphasize early control of disease activity, prevention of organ damage, appropriate use of hydroxychloroquine and immunosuppressive therapy, and minimization of glucocorticoid exposure.[15] The high mortality observed in our cohort highlights the importance of early diagnosis, prompt treatment of severe organ involvement, and close follow-up of children presenting with highly active disease.

CONCLUSION

Childhood-onset SLE showed a marked female predominance and commonly presented with serosal, hematological, neurological, and renal involvement. Most children had high SLEDAI-2K disease activity at diagnosis, while Class IV lupus nephritis was the predominant renal histopathological pattern. Although disease activity markedly improved among survivors at six months, the high early mortality emphasizes the need for early recognition, aggressive management of severe disease, and close follow-up.

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