Performance of 2019 EULAR/ACR Classification Criteria for Systemic Lupus Erythematosus in Children and Adults : A Retrospective Observational Study

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Abstract

Background: The European League Against Rheumatism (EULAR) and American College of Rheumatology (ACR) recently developed a systemic lupus erythematosus (SLE) classification criteria (EULAR/ACR-2019) with high sensitivity and specificity. The aim of this study was to validate and compare the performance of the newly developed criteria to that of the ACR-1997 and the 2012 Systemic Lupus International Collaborating Clinics (SLICC-2012) criteria in juvenile-onset SLE (jSLE) and adult-onset SLE (aSLE) patients.

Methods: We conducted a retrospective study of SLE patients (221 children and adult) and controls (214 children and adult) with defined rheumatic diseases to establish the ACR-1997, SLICC-2012 and EULAR/ACR-2019 criteria. The performance of the three criteria was statistically analyzed.

Results: For jSLE, sensitivities of ACR-1997, SLICC-2012 and EULAR/ACR-2019 criteria were 63.3%, 94.6% and 98.2% ($P < 0.001$), with specificities 99.5%, 98.6% and 93.5% ($P < 0.001$), respectively. For aSLE, sensitivities of ACR-1997, SLICC-2012 and EULAR/ACR-2019 criteria were 72.9%, 96.8% and 99.1% ($P < 0.001$), with specificities 97.2%, 92.5% and 90.2% ($P = 0.013$), respectively. In ANA positive juvenile patients, a EULAR/ACR score ≥ 13 instead of a score ≥ 10 resulted in higher specificity (93.1% vs. 75.9%), despite slightly lower sensitivity (92.2% vs. 99.5%). In both jSLE and aSLE patients, the SLICC-2012 and EULAR/ACR-2019 criteria had increased sensitivity for major organ involvement than ACR-1997.

Conclusion: The EULAR/ACR-2019 criteria showed similar sensitivity to jSLE and aSLE patients and was more sensitive than ACR-1997 and SLICC-2012 criteria, allowing earlier recognition of patients with single or major organ involvement. The adoption of a EULAR/ACR total score ≥ 13 in this study, instead of the initially proposed ≥ 10 score, was more appropriate to classify jSLE.

Background

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by the production of autoantibodies and multisystemic involvement. Although it affects mainly women of child-bearing age, disease onset can occur in children and teenagers. The heterogeneity of disease manifestations and pathogenesis has posed a diagnostic challenge for clinicians. Juvenile-onset SLE (jSLE), defined as the onset of SLE in patients before 18 years of age, is different from adult-onset SLE (aSLE) in terms of disease pattern [1]. The differences in clinical manifestations and laboratory features in jSLE and aSLE attribute to the distinct diagnostic approach in clinical scenario.

No diagnostic criteria have been proposed in SLE. However, classification criteria are important for the identification of relatively homogeneous patients for inclusion in clinical research and trials. To date, the widely used classification criteria for SLE include the 1982 revised American College of Rheumatology (ACR-1982) criteria [2], the 1997 updated ACR criteria (ACR-1997) [3], and the 2012 Systemic Lupus International Collaborating Clinics (SLICC-2012) criteria [4]. These classification criteria were developed and validated mainly in aSLE patients. Limited numbers of studies have validated the performance of these criteria in jSLE patients [5-8]. Most recently, in 2019, the EULAR (European League Against Rheumatism) and ACR jointly initiated a four-phase project to develop more sensitive and specific SLE classification criteria (EULAR/ACR-2019). In the criteria set, ANA has been defined as the required entry criterion. There are seven clinical and three immunological domains, and the patient is classified with SLE when the total score ≥ 10 [9]. The new criteria aimed to improve sensitivity and promote earlier identification of lupus. Although the new criteria had excellent sensitivity and specificity, it was not widely validated in special subgroups of SLE including pediatric patients, different racial/ethnic groups [9]. There're currently six studies describing the performance of EULAR/ACR-2019 criteria in jSLE patients with various conclusions [10-15]. However, no studies have validated and compare the performance of the newly developed criteria in both jSLE and aSLE in Chinese population from the same medical center.

This study aims to compare the performance of ACR-1997, SLICC-2012 and the new EULAR/ACR-2019 criteria in a large cohort of Chinese jSLE and aSLE patients from the same medical center, so as to validate the performance of EULAR/ACR-2019 classification criteria in this group of SLE patients.

Methods

Study Population

We performed a retrospective cross-sectional study in the Department of Pediatrics and Rheumatology, Renji Hospital, Shanghai Jiao Tong University. 221 jSLE patients diagnosed in Renji Hospital from January 2013 to October 2020 were continuously enrolled in the study. 221 randomly selected aSLE patients diagnosed at the same period in the Department of Rheumatology were enrolled. The control groups consisted of 214 patients with other defined rheumatic diseases in children and adult respectively. Controls were selected due to their similarity with SLE, which included patients diagnosed with the following connective tissue diseases: juvenile idiopathic arthritis, vasculitis, dermatomyositis, undifferentiated connective tissue disease, Behcet's disease, mixed connective tissue disease, scleroderma, Sjogren's syndrome, rheumatoid arthritis, adult onset still disease. All the patients' diagnosis were confirmed by experienced rheumatologists. We used the physician's diagnosis of SLE as the gold standard.

Demographic, clinical, and laboratory features, including the items of different criteria sets [3, 4, 9] were evaluated through chart review. Two experienced rheumatologists independently evaluated each case according to the three sets of SLE criteria (ACR-1997, SLICC-2012 and EULAR/ACR-2019) [3, 4, 9]. Cases of disagreement were discussed until consensus were reached. ANA test result was defined as positive if staining reactivity was positive at $\geq 1:80$ serum dilution. Disease activity was assessed by SLEDAI-2K (Systemic Lupus Erythematosus Disease Activity Score, version 2000) as described elsewhere [16]. The sensitivity and specificity of the criteria sets were evaluated based on the features of the patients at the time of disease diagnosis.

The protocol of this study was approved by the Ethics Committee of Renji Hospital, Shanghai Jiao Tong University, School of Medicine. Informed consents were obtained from all parents/patients before inclusion in the study.

Statistical analysis

The SPSS version 26.0 (SPSS, Inc., Chicago, USA) and GraphPad Prism version 9.1.1 software (GraphPad Software, La Jolla, CA, USA) were used for statistical analysis. All data are expressed using mean and standard deviation (SD) or frequencies and percentages. Continuous variables were compared between groups using student's t test or Mann-Whitney u test. Categorical variables were compared using the Chi-square test or Fisher's exact test. Sensitivity, specificity, accuracy, positive and negative likelihood and diagnostic odds ratio along with confidence intervals, were performed for each set of criteria. McNemar's test was performed to assess differences in sensitivity and specificity between ACR-1997, SLICC-2012 and EULAR/ACR-2019 criteria. We used the Receiver Operating Characteristics (ROC) curve to demonstrate the best performing cut-off value for the EULAR/ACR-2019 criteria in our study group (only ANA positive patients were included). Pearson correlation analysis was used to analyze the correlation between variables. $P < 0.05$ was considered statistically significant, and the confidence interval was 95%.

Results

Sample demographics

Demographic characteristics of the cohort are presented in Table 1. For juvenile patients, the study population included 221 continuous cases with a well-established clinical diagnosis of jSLE (83.7% female, a mean age at diagnosis of 10.9 ± 2.7 years) and 214 controls. The jSLE group had a higher female ratio ($P < 0.001$) and older diagnosis age ($P < 0.001$) than the controls. In adult population, 221 cases with aSLE (90.5% female, a mean age at diagnosis of 36.8 ± 12.8 years) and 214 controls were enrolled in this study. The aSLE had a higher female ratio ($P < 0.001$) and younger diagnosis age ($P < 0.001$) than the controls. Both jSLE and aSLE group had a higher proportion of patients with positive ANA than the control group. The control groups included a variety of diseases detailed in Table 1. In juvenile patients, juvenile idiopathic arthritis ($n=90$) and vasculitis ($n=44$) were the most common. In adult patients, the most prevalent diagnoses were dermatomyositis ($n=30$) and vasculitis ($n=30$).

Performance of the criteria

Performance of EULAR/ACR-2019 criteria, SLICC-2012, and ACR-1997 are presented in Table 2. In jSLE patients, the sensitivity of the EULAR/ACR-2019 criteria was the highest (98.2%), while the highest specificity was that of the ACR-1997 criteria (99.5%). The SLICC-2012 criteria performed well with high sensitivity and specificity (94.6% and 98.6%). For aSLE, both the EULAR/ACR-2019 and SLICC-2012 criteria had higher sensitivity (99.1% and 96.8%, respectively) than the ACR-1997 (72.9%), while the ACR-1997 criteria had higher specificity than the other two sets (97.2% vs. 90.2%–92.5%).

Performance of the criteria in ANA positive patients

Since a positive ANA is the entry criteria in EULAR/ACR-2019 criteria, we also evaluated the performances of the three criteria sets by including only ANA positive SLE patients and controls (Table 2). There were three ANA negative patients in our jSLE cohort, the sensitivities of the ACR-1997, SLICC-2012 and EULAR/ACR-2019 criteria were 64.2%, 94.5%, and 99.5%, respectively; while the specificities were 98.3%, 94.8%, and 75.9%, respectively. For adult patients, the sensitivities of the ACR-1997, SLICC-2012 and EULAR/ACR-2019 criteria were 72.9%, 96.8%, and 99.1%, respectively. The specificities in aSLE patients were 95.7%, 88.7%, and 85.1%, respectively. When including only ANA positive SLE patients and controls, we noticed a difference in specificities by the EULAR/ACR-2019 criteria. Especially in jSLE patients, the specificity of EULAR/ACR-2019 criteria decreased from 93.5% (all patients) to 75.9% (including only ANA positive patients).

Receiver operating characteristic (ROC) analysis showed a good performance of EULAR/ACR-2019 criteria in discriminating SLE from other rheumatic disease patients (Figure 1). Among ANA positive juvenile patients, the area under the ROC curve for EULAR/ACR-2019 criteria was 0.976 (95%CI, 0.959-0.994), and the cut-off value was 13, resulting in a sensitivity and a specificity of 92.2% and 93.1%, respectively. When we picked ≥ 13 as the threshold for the EULAR/ACR-2019 criteria, the sensitivity slightly decreased (from 99.5% to 92.2%), but the specificity significantly increased (from 75.9% to 93.1%). For adult patients, the area under the ROC curve for EULAR/ACR-2019 criteria was 0.965 (95% CI, 0.946-0.985), and the cut-off value was 10 (sensitivity, 99.1%; specificity, 85.1%).

Proportion of patients meeting overlapping or individual criteria

Among the clinically diagnosed SLE patients, 140 (63.3%) jSLE and 161 (72.9%) aSLE patients met all three classification criteria. There were twelve jSLE patients and seven aSLE patients who only met the EULAR/ACR-2019 criteria, of whom eleven and four cases had single organ involvement, respectively. Four cases of jSLE patients were missed by the EULAR/ACR-2019 criteria, three of whom were ANA negative (one turned ANA positive three months after diagnosis). Two aSLE patients were missed by the EULAR/ACR-2019 criteria, both were ANA positive, and one patient got a EULAR/ACR total score of eight (kidney biopsy showed V-type lupus nephritis) (Figure 2). Fourteen children and twenty-one adult patients in controls were misclassified as SLE by the EULAR/ACR-2019 criteria. Patients with undifferentiated connective tissue disease (UCTD) and mixed connective tissue disease (MCTD) were most likely to be classified as SLE by the EULAR/ACR-2019 criteria in our cohort.

Sensitivity of the criteria across subgroups of SLE patients with major organ involvement

SLE patients with major organ involvement often accrue more damage, which has negative impact on long term survival. The prompt diagnosis and management is essential to improve patients' prognosis. Thus, we specifically analyzed the diagnostic performance of the three criteria in patients with major organ involvement. Compared with the other two classification criteria, ACR-1997 had lower sensitivity for classifying neurological, renal, and hematological SLE in both children and adults. The SLICC-2012 and EULAR/ACR-2019 criteria had increased sensitivity for hematological manifestations. In jSLE patients, the sensitivity of the SLICC-2012 and EULAR/ACR-2019 criteria were the same (97.6%), while in aSLE, the sensitivity of EULAR/ACR-2019 was slightly higher (99.3% vs. 97.9%). The sensitivity of the SLICC-2012 for classifying renal jSLE was slightly higher than EULAR/ACR-2019 criteria (100% vs. 96.9%), whereas in

aSLE, the sensitivity of the two criteria sets was the same (99.2%). The three criteria set had high sensitivity for classifying SLE patients with moderate and severe activity (SLEDAI-2K ≥ 10) (Figure 3).

jSLE patients fulfilling the EULAR/ACR-2019 criteria had a mean score of 19.3 (range:10-33). SLEDAI-2K ranged from 3 to 26 (mean, 10.2). aSLE patients who met the EULAR/ACR-2019 criteria scored 10-41 (mean, 20.8). SLEDAI-2K ranged from 2 to 27 (mean, 12.3). There was a positive correlation between EULAR/ACR total score and SLEDAI in jSLE and aSLE groups ($r=0.700$, $P<0.001$; $r=0.748$, $P<0.001$) (Figure 4).

Discussion

SLE is a complex autoimmune disease with diverse clinical manifestations. There is no gold standard for the diagnosis of SLE up to now. The classification criteria of SLE have always been the basis of clinical studies. Our results demonstrate that the EULAR/ACR-2019 criteria showed improved sensitivity than the other two criteria and promoted earlier identification of major organ involved lupus. By adjusting the classification thresholds, the specificity of the new criteria for classifying SLE is enhanced. To the best of our knowledge, this is the first study assessing the performance of the three classification systems (ACR-1997, SLICC and especially the new EULAR/ACR criteria) applied to both jSLE and aSLE patients in Chinese population from the same medical center.

Several studies have compared the performances of these three criteria sets. Batu et al. [10] found that SLICC-2012 yielded the best sensitivity (95.4%) and was more suitable for jSLE patients than EULAR/ACR-2019 criteria. A meta-analysis conducted by Hartman et al. showed that for jSLE patients, SLICC-2012 criteria had higher sensitivity (99.9% vs. 84.3%) than ACR-1997, but much lower specificity (82% vs. 94.1%), so ACR-1997 criteria was more suitable for jSLE patients [5]. Aljaberi et al. [11] compared the criteria sets in a cohort of 112 SLE patients aged 2-21 years and 105 controls, they demonstrated that the EULAR/ACR-2019 criteria had significantly higher sensitivity (85% vs. 72%) and similar specificity (83% vs. 87%) than the ACR-1997 criteria. In aSLE patients, Dahlstrom et al. [17] found that the EULAR/ACR-2019 and SLICC-2012 criteria had higher sensitivity (93% and 100%, respectively) than ACR-1997 (83%), but lower specificity than ACR-1997, corresponding to the results of our study. While another study in Japan indicated that the EULAR/ACR criteria accomplished modestly high sensitivity (92%), but not as high as the ACR-1997 and SLICC-2012 criteria (97% and 99%, respectively), probably due to the cut-off value of ANA titer [18].

ANA testing is widely used for SLE diagnosis, based on its high sensitivity [19]. In our study, the frequency of ANA in aSLE patients was 100%, higher than that of jSLE patients (98.6%), which is similar to the estimates of a large meta-regression of epidemiological studies (97.8%, 95%CI: 96.8% to 98.5%) [20]. The EULAR/ACR-2019 criteria may miss a significant proportion of jSLE patients, mostly because of the absence of ANA positivity. On the other hand, positive ANA can be found in a variety of autoimmune diseases, which may result in misclassification as SLE based on the new criteria [14]. Within ANA-positive jSLE patients, the sensitivity of the EULAR/ACR criteria was higher (99.5%), but the specificity dropped to 75.9%, significantly lower than the results (98.4%) of Ma et al. [12]. To overcome this, we performed ROC analysis in ANA positive subjects and found that EULAR/ACR total score cut-off point influenced its performance measures. When a EULAR/ACR score ≥ 13 was selected as the cut-off point, determined by ROC curve analysis, the sensitivity (from 99.5% to 92.2%) was slightly decreased, but the specificity (from 75.9% to 93.1%) was significantly improved compared with the initially proposed ≥ 10 score, consistent with the study of Rodrigues et al. [13].

Differences between our results with those in other cohorts might be due to various factors such as patients' demographics and inclusion of disease controls. Patients with UCTD and MCTD in controls of our study were most likely to be classified as having SLE by the EULAR/ACR-2019 criteria, due to the high weight of arthritis and mucocutaneous manifestations. A patient gets six out of the required 10 points from only arthritis according to the EULAR/ACR-2019 criteria. However, in the EULAR/ACR criteria, a general attribution rule has been defined as counting an item in favor of SLE only if SLE is the most likely explanation, so all clinical and immunologic manifestations should be considered comprehensively. The study of Radin et al. [21] included 133 patients with UCTD, of whom 22 (17%) cases fulfilled the EULAR/ACR-2019 criteria. In another study, when applying the SLICC-2012 and EULAR/ACR criteria, 18 (14.0%) and 26 (20.2%) patients with UCTD were reclassified as SLE [22]. The new criteria will help to early identify patients at higher risk of developing more severe CTD manifestations. On the other hand, specificity estimates for criteria sets depend on the choice of the control population.

Patients with major organ involvement often need immunosuppressive therapy. Prompt treatment is critical to ensure good outcomes in this subgroup of patients. Early diagnosis of such patients with SLE is of great significance [23]. Adamichou et al. [24] showed that all three criteria had lower sensitivity for neurological than renal SLE. In both jSLE and aSLE patients of our cohort, the ACR-1997 criteria had relatively low sensitivity to neurological, renal, and hematological SLE, while the SLICC-2012 and EULAR/ACR criteria had pretty high sensitivity to hematological and renal SLE. The SLICC-2012 criteria divide hematological abnormalities into three independent parts. Thus, a patient could meet three out of the required four criteria with only hematologic involvement according to the SLICC-2012 criteria [4]. These hematological abnormalities were weighted by the EULAR/ACR criteria, and the highest rank a patient could get from this domain is four which occurs in the presence of autoimmune hemolysis or thrombocytopenia. In this case, along with a positive anti-dsDNA or anti-Sm antibodies, a patient could be classified as SLE [9]. This might have contributed significantly to the higher sensitivity of the two criteria sets to hematological SLE.

The emergence of organ-dominant forms poses further challenges to classification and diagnosis. The SLICC-2012 first introduced the renal stand-alone criterion for classifying renal-dominant lupus, and the EULAR/ACR introduced higher-weighted items to enable classification with fewer number of afflicted organs. In the EULAR/ACR criteria, class III or IV lupus nephritis confirmed by renal biopsy has the highest weight and can be classified as SLE by its mere presence. Class II or V lupus nephritis also has a high weight but is not sufficient by its mere presence. It supports the idea of SLICC-2012 and is more reasonable than SLICC-2012.

This study has some limitations. Firstly, the retrospective design and data extraction from medical records may lead to the omission of some important clinical information, although the prevalence of various single criteria indicate this is not a major problem. Secondly, we were not able to compare the

performances of different criteria sets according to the disease course and follow-up. Finally, the study was conducted in only a single center, a larger cohort of patients and multicenter studies are expected to fully validate the performance of the 2019 EULAR/ACR criteria in the Chinese SLE population.

Conclusion

In conclusion, the EULAR/ACR-2019 criteria exhibited high sensitivity in jSLE and aSLE patients, allowing earlier recognition of patients with single or major organ damage. However, the newly established criteria may result in over diagnosis for mimic phenotype was more prevalent in children and an improved threshold score may be more appropriate for paediatric population. Meanwhile, since the EULAR/ACR remains a classification criterion rather than a diagnostic criteria, patients who do not meet the classification criteria should not be prevented from receiving timely and appropriate treatment measures.

Abbreviations

SLE: systemic lupus erythematosus; jSLE: juvenile-onset SLE; aSLE: adult-onset SLE; SD: standard deviation; ANA: anti-nuclear antibody; UCTD: undifferentiated connective tissue disease; MCTD: mixed connective tissue disease; ANA⁺: anti-nuclear antibody positive; PPV: positive predictive value; NPV: negative predictive value; EULAR: European League Against Rheumatism; ACR: American College of Rheumatology; SLICC: Systemic Lupus International Collaborating Clinics; CI: confidence interval; AUC: area under curve; ROC: receiver operating characteristic; SLEDAI-2K: systemic lupus erythematosus disease activity index 2000.

Declarations

Acknowledgements

Not applicable.

Authors' contributions

CL conceived of the study and participated in its design and coordination. CS, DH, and XH contributed to the data acquisition and analysis. TZ, XM and ZW made critical review for intellectual content. CS and DH drafted the manuscript and revised the manuscript. All authors read, revised, and approved the final manuscript.

Authors' information

Sang Cheng and Huihua Ding contributed equally to this article.

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Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Ethics approval and consent to participate

The protocol of this study was approved by the Ethics Committee of Renji Hospital, Shanghai Jiao Tong University, School of Medicine.

Consent for publication

Not applicable.

Competing interests

The authors declare that they have no competing interests.

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Tables

Table 1. Demographic characteristics of study population

Characteristics	jSLE (n=221)	Juvenile controls (n=214)	<i>P</i> -value	aSLE (n=221)	Adult controls (n=214)	<i>P</i> -value
Female, n (%)	185(83.7)	114(53.3)	<0.001	200(90.5)	151(70.6)	<0.001
Age at diagnosis, mean ± SD, years	10.9±2.7	8.1±3.5	<0.001	36.8±12.8	51.2±16.0	<0.001
ANA positive, n (%)	218(98.6)	58(27.1)	<0.001	221(100)	141(65.9)	<0.001
Controls, disease; n						
Juvenile idiopathic arthritis		90				
Vasculitis		44			30	
Dermatomyositis		30			30	
UCTD		25			24	
Behcet's disease		9			10	
MCTD		8			21	
Scleroderma		8			14	
Sjogren's syndrome					25	
Rheumatoid arthritis					20	
Adult onset still disease					30	

SLE: systemic lupus erythematosus; jSLE: juvenile-onset SLE; aSLE: adult-onset SLE; SD: standard deviation; ANA: anti-nuclear antibody; UCTD: undifferentiated connective tissue disease; MCTD: mixed connective tissue disease.

Table 2. Performance measures for the three classification criteria in jSLE and aSLE patients (with all patients and ANA positive patients, respectively)

	ACR-1997		SLICC-2012		EULAR/ACR-2019		ACR-1997		SLICC-2012		EULAR/ACR-2019	
	jSLE	ANA ⁺ jSLE	jSLE	ANA ⁺ jSLE	jSLE	ANA ⁺ jSLE	aSLE	ANA ⁺ aSLE	aSLE	ANA ⁺ aSLE	aSLE	ANA ⁺ aSLE
Sensitivity	0.633	0.642	0.946	0.945	0.982	0.995	0.729	0.729	0.968	0.968	0.991	0.991
(95% CI)	(0.569-0.698)	(0.578-0.706)	(0.916-0.976)	(0.914-0.975)	(0.964-1.000)	(0.986-1.004)	(0.669-0.788)	(0.669-0.788)	(0.945-0.992)	(0.945-0.992)	(0.978-1.004)	(0.978-1.004)
Specificity	0.995	0.983	0.986	0.948	0.935	0.759	0.972	0.957	0.925	0.887	0.902	0.851
(95% CI)	(0.986-1.005)	(0.948-1.017)	(0.970-1.002)	(0.890-1.007)	(0.901-0.968)	(0.645-0.872)	(0.950-0.994)	(0.924-0.991)	(0.890-0.961)	(0.834-0.940)	(0.862-0.942)	(0.792-0.911)
PPV(%)	99.3	99.3	98.6	98.6	93.9	93.9	96.4	95.8	93.0	93.0	91.3	91.3
NPV(%)	72.4	42.2	94.6	82.1	98.0	97.8	77.6	69.2	96.6	94.7	99.0	98.4
Accuracy	0.811	0.714	0.966	0.946	0.959	0.946	0.848	0.818	0.947	0.936	0.947	0.936
(95% CI)	(0.775-0.848)	(0.660-0.767)	(0.948-0.983)	(0.919-0.973)	(0.940-0.977)	(0.919-0.973)	(0.814-0.882)	(0.778-0.858)	(0.926-0.968)	(0.911-0.962)	(0.926-0.968)	(0.911-0.962)

ANA⁺: anti-nuclear antibody-positive; SLE: systemic lupus erythematosus; jSLE: juvenile-onset SLE; aSLE: adult-onset SLE; PPV: positive predictive value; NPV: negative predictive value; EULAR: European League Against Rheumatism; ACR: American College of Rheumatology; SLICC: Systemic Lupus International Collaborating Clinics; CI: confidence interval.

Figures

Figure 1

ROC curve of the EULAR/ACR-2019 criteria in ANA-positive SLE patients and controls. The area under the ROC curve and the cut-off value of EULAR/ACR-2019 criteria in jSLE (A) and aSLE (B) patients. SLE: systemic lupus erythematosus; jSLE: juvenile-onset SLE; aSLE: adult-onset SLE; EULAR: European League Against Rheumatism; ACR: American College of Rheumatology; ANA: anti-nuclear antibody; AUC: area under curve; ROC: receiver operating characteristic; CI: confidence interval.

Figure 2

Venn diagram illustrating proportion of patients meeting overlapping or individual criteria in jSLE and aSLE cohort. Proportion of patients meeting EULAR/ACR-2019, SLICC-2012 and ACR-1997 criteria in jSLE (A) and aSLE (B) cohort. SLE: systemic lupus erythematosus; jSLE: juvenile-onset SLE; aSLE: adult-onset SLE; EULAR: European League Against Rheumatism; ACR: American College of Rheumatology; SLICC: Systemic Lupus International Collaborating Clinics.

Figure 3

Performance of the three criteria in SLE patients with major organ involvement and moderate/severe disease activity. Sensitivity of EULAR/ACR-2019, SLICC-2012 and ACR-1997 criteria across subgroups of jSLE (A) and aSLE (B) patients with neurological, hematological, renal involvement and moderate/severe disease activity. Moderate/severe disease activity was defined as SLEDAI-2K ≥ 10 . SLE: systemic lupus erythematosus; jSLE: juvenile-onset SLE; aSLE: adult-onset SLE; EULAR: European League Against Rheumatism; ACR: American College of Rheumatology; SLICC: Systemic Lupus International Collaborating Clinics.

Figure 4

The correlation of EULAR/ACR-2019 criteria with SLEDAI in juvenile and adult SLE patients. EULAR/ACR-2019 criteria correlated positively with SLEDAI in both jSLE (A) and aSLE (B) patients. SLE: systemic lupus erythematosus; jSLE: juvenile-onset SLE; aSLE: adult-onset SLE; EULAR: European League Against Rheumatism; ACR: American College of Rheumatology; SLEDAI: systemic lupus erythematosus disease activity index.