Performance of 2019 EULAR/ACR classification criteria for Systemic Lupus Erythematosus in a pediatric population – a multicenter study

Yoel Levinsky[1,2], Mor Broide[2,3], Shelly Kagan[1,2], Ori Goldberg[2,4], Oded Scheuerman[1,2], Rotem Tal[2,5], Irit Tirosh[2,6], Yonatan Butbul Aviel[7,8], Liora Harel[2,5], Gil Amarilyo[2,5]

[1] Department of Pediatrics B, Schneider Children's Medical Center of Israel, Petach Tikva, Israel; [2] Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel; [3] Department of Pediatrics A, Schneider Children's Medical Center, Petah Tikva, Israel [4] Pediatric Pulmonary Unit, Schneider Children's Medical Center of Israel, Petach Tikva, Israel [5] Pediatric Rheumatology Unit, Schneider Children's Medical Center of Israel, Petach Tikva, Israel; [6]Pediatric Rheumatology service, Edmond and Lily Safra Children's Hospital, Sheba Medical Center, Tel Hashomer ; [7] Pediatric Rheumatology Service, Ruth Rappaport Children's Hospital, Rambam Health Care Campus, Haifa, Israel; [8]The Ruth and Bruce Rappaport Faculty of Medicine, Technion - Israel Institute of Technology, Haifa, Israel

Abstract

Background: The "European League Against Rheumatism" and "American College of Rheumatology" 2019 (EULAR/ACR-19) criteria for the diagnosis of Systemic Lupus Erythematosus (SLE) were recently published, with the stated goal of maintaining the level of sensitivity and raising the level of specificity for classification of SLE in adults.

Objectives: The aim of this study is to examine the EULAR/ACR-19 criteria application to juvenile SLE (jSLE) patients.

Methods: In this multicenter study the charts of jSLE patients from three tertiary medical centers were reviewed and compared to patients with non-jSLE diagnosis. Pediatric rheumatologists, blinded to the original diagnosis, reviewed and diagnosed all cases. Pediatric patients' clinical and laboratory data were retrospectively extracted and then examined with regard to how they met the new and old criteria.

Results: Included were 225 patients (112 jSLE, 113 non-SLE). When applied to juvenile SLE classification, the sensitivity of the new EULAR/ACR-19 criteria was 0.96 (0.9-.0.99) and the specificity was 0.89 (0.82-0.94). These were comparable to the Systemic Lupus International Collaborating Clinics (SLICC) criteria. . The sensitivity of the EULAR/ACR-19 criteria improves over time and was 0.83 twelve months following disease onset, reaching 0.96 after longer than 24 months.

Conclusion: Among a cohort of jSLE patients, sensitivity of the new EULAR/ACR-19 criteria was found to be high and specificity may have improved slightly compared to the SLICC-12 criteria. We support the use of the new classification criteria for

pediatric patients in future jSLE studies, but it should be noted that its specificity is lower than for adults.

	ACR-97	SLICC-12	EULAR/ACR-19
Sensitivity (95% CI)	0.79 (0.70-0.86)	0.96 (0.9-0.99)	0.96 (0.9-.0.99)
Specificity (95% CI)	0.94 (0.88-0.97)	0.85 (0.77-0.91)	0.89 (0.82-0.94)
Accuracy (95% CI)	0.86 (0.81-0.9)	0.9 (0.86-0.94)	0.92 (0.88-.0.96)
Positive Likelihood Ratio (95% CI)	12.7(6.1-26.2)	6.35(4.1-9.9)	9.0 (5.3-15.4)
Negative Likelihood Ratio (95% CI)	0.23(0.16-0.33)	0.05(0.02-0.12)	0.05(0.02-0.12)
Diagnostic odds ratio (95% CI)	55.5(22.80-135.0)	120.85(43.0-340.0)	180.1(61.3-529.4)

Receiver operating characteristics (ROC) curve, comparing performance of three sets of criteria for the diagnosis of juvenile-onset systemic lupus erythematosus