Correspondence on 'Lupus or not? SLE Risk Probability Index (SLERPI): a simple, clinician-friendly machine-learning-based model to assist the diagnosis of systemic lupus erythematosus'

We read the article by Adamichou et al, 'Lupus or not? SLE Risk Probability Index (SLERPI): a simple, clinician-friendly machine-learning-based model to assist the diagnosis of systemic lupus erythematosus' with great interest.[1] They proposed a machine-learning-based index (Systemic Lupus Erythematosus Risk Probability Index (SLERPI)) with 14 weighted items to predict the diagnosis of SLE. The original model gives SLE risk probabilities as definite SLE (87%–100%), likely SLE (44%–86%), possible/cannot rule out SLE (15%–43%) and unlikely SLE (0%–14%). The authors have also converted their model into a simple scoring system to facilitate its use in daily practice. Using their model on binary (SLE if score >7 and not SLE if ≤7), they reported a sensitivity of 94.2% and a specificity of 94.4% in their cohort. Since the original cohort was based on adult patients, we have tested the performance of the binary model in our paediatric SLE cohort.

We included 111 paediatric SLE patients and 171 controls with other rheumatologic diseases (all diagnosed before 18 years of age) followed at the Department of Pediatric Rheumatology in our university hospital. These patients have also been included in our recent paper testing the performances of the existing SLE classification criteria in paediatric SLE.[2] The diagnosis of SLE was based on expert opinion. In the control group, the most prevalent diagnoses were primary systemic vasculitis and juvenile dermatomyositis. Patient and control data were collected on standardised case report forms. The sensitivity and specificity of the SLERPI binary model was evaluated based on the features of the patients at the time of disease diagnosis.

The sensitivity and the specificity of the SLERPI binary model (with a cut-off value of 7) was 90% and 81.2%, respectively, in our cohort. The area under the receiver operating characteristic curve was 0.94 (figure 1) which indicates a good discrimination (SE: 0.012; 95% CI: 0.919 to 0.968). When we chose >8 as threshold, the sensitivity slightly decreased (from 90% to 88.2%) while the specificity increased (from 81.2% to 89.4%). Thus, a threshold of 8 worked better in our cohort. Haematologic abnormalities such as thrombocytopenia or haemolytic anaemia and nephritis are more frequent among childhood patients than adults.[3] Both lupus nephritis and haematologic features of SLE are the highest weighted items in the binary SLERPI model. This may be the reason for a higher threshold to perform better in the paediatric SLE patients.

Eleven SLE patients had a ≤7 score from SLERPI and were not classified with SLE. None of these patients had lupus nephritis or haematologic involvement. When we analysed the characteristics of controls misclassified with SLE by SLERPI (n=32), the most common diagnoses were haemolytic uremic syndrome (HUS) and mixed connective tissue disease (MCTD). Patients with MCTD had similar clinical features with SLE and the differential diagnosis is challenging between MCTD and SLE in clinical practice. On the other hand, the haematologic manifestations that are common between HUS and SLE, which are highly weighted in the SLERPI model, were probably the main reasons for HUS misclassification as SLE. When we raised the threshold from 7 to 8, only 18 out of these 32 patients (56.3%) were misclassified with SLE. It was noteworthy that patients with MCTD and HUS were again misclassified with SLE by SLERPI model with the threshold of 8.

In conclusion, with the threshold set for adults, the SLERPI binary model yielded a low sensitivity and specificity in paediatric SLE cohort. However, this model can be useful in the paediatric practice when the threshold is set at >8.

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Contributors All authors were involved in drafting and revising the manuscript, and approved the final version. Study design: EDB, YB and SO. Data collection: EDB, UKA, YB and OB. Interpretation and analysis of the data: EDB, UKA, OB and SO.

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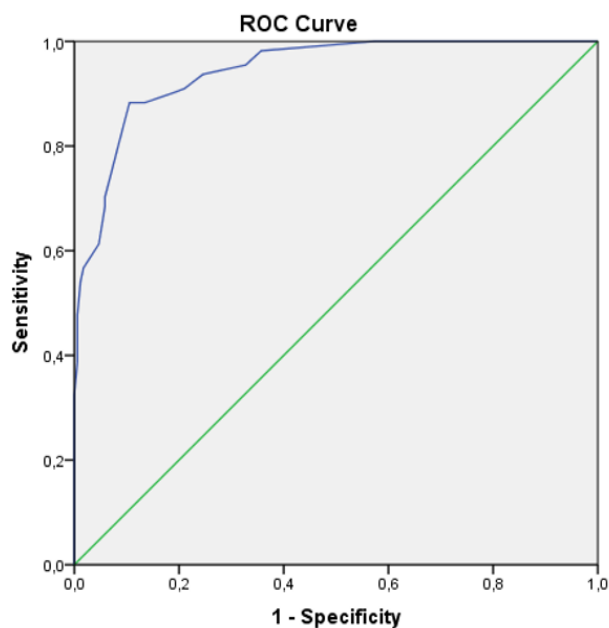
Figure 1 Area under the receiver operating characteristic (ROC) curve for the Systemic Lupus Erythematosus (SLE) Risk Probability Index binary model in paediatric SLE patients and controls.