

## 2019 European League Against Rheumatism/American College of Rheumatology Classification Criteria for Systemic Lupus Erythematosus

Supplement 2: Patient examples of correctly using the EULAR/ACR 2019 classification criteria

- 1) 30 year female patient with new onset polyarthritis. Anti-CCP antibodies negative, no erosions on plain x-ray films, HEp-2 ANA 1:320 (homogenous), anti-dsDNA antibodies positive in ELISA and verified by CLIFT (Crithidia luciliae immunofluorescence test)

Entry criterion: Fulfilled (**ANA ever positive** by HEp-2  $\geq$ 1:80 or equivalent test)

Clinical criterion: Fulfilled (Joint involvement): **6 points**

Attribution to SLE: Correct (Anti-CCP is negative, no more likely cause of arthritis)

Lupus specific antibody: anti-dsDNA positive (CLIFT has >90% proven specificity against relevant disease controls, ELISA anti-dsDNA alone usually not): **6 points**

**ANA entry + 12 points: This patient should be classified as having SLE.**

- 2) 20-year old female patient with fever, oral ulcers, leukocytopenia and thrombocytopenia. The patient is ANA positive (Labcorp result). Specific antibodies are negative, complement levels are normal

Entry criterion: Fulfilled (**ANA ever positive** by HEp-2  $\geq$ 1:80 or equivalent test)

Clinical criterion: Fulfilled (Fever, oral ulcers, leukopenia, thrombocytopenia)

Attribution to SLE: All questionable (count only if no more likely explanation than SLE)

If attribution of all correct

- Hematology: Thrombocytopenia and leukopenia in same domain (count only highest number): **4 points**
- Oral ulcers: **2 points**
- Fever: **2 points**

**ANA entry + 8 points: This patient should NOT be classified as having SLE**, even if there is no more likely reason for fever, oral ulcers and combined leukopenia/thrombocytopenia (such as virus infection, leukemia).

- 3) 25 year old female patient with deep vein thrombosis, pulmonary embolism, pleural effusion, fever, and thrombocytopenia. HEp-2 ANA 1:160 (speckled), anti-phospholipid antibodies triple-positive, specific SLE antibodies negative, complement levels normal.

Entry criterion: Fulfilled (**ANA ever positive** by HEp-2  $\geq 1:80$  or equivalent test)

Clinical criterion: Fulfilled (Fever, thrombocytopenia)

Attribution to SLE: No - for thrombocytopenia and anti-phospholipid antibodies primary APS more likely explanation than SLE, for pleural effusion and fever pulmonary embolism more likely explanation than SLE.

**ANA entry + 0 points** (neither 2 points for fever nor 5 points for pleural effusion or 4 points for thrombocytopenia to be counted because of more likely attribution to another cause than SLE): **This patient should NOT be classified as having SLE.**

- 4) 45 year old female patient with biopsy proven lupus nephritis ISN/RPS class IV, treated with EuroLupus cyclophosphamide. Proteinuria still 1.2 g/day (from 3.4 g/day). ANA positive at diagnosis, but now negative. Anti-dsDNA ELISA positive at diagnosis, CLIFT now negative.

Entry criterion: Fulfilled (**ANA ever positive** HEp-2  $\geq 1:80$  or equivalent test)

Clinical criterion: Fulfilled (Nephritis)

Nephritis: Histology and proteinuria in same domain (count only highest number): **10 points**

Anti-dsDNA: Do not count ELISA (unless specificity of  $\geq 90\%$  against relevant disease controls proven), even though CLIFT or other specific test might have been positive at diagnosis.

**ANA entry (historical sufficient) + 10 points: This patient should be classified as having SLE.**