Background The 1997 Update of the 1982 American College of Rheumatology Revised Criteria for Classification of SLE includes two autoantibody criteria: #10, abnormal level of anti-native DNA, anti-Sm, or antiphospholipid; #11 positive antinuclear antibody (ANA). Thus, ANA positivity is counted as 1 of the 11 criteria and a person shall be said to have SLE if any 4 or more of the 11 criteria are present, serially or simultaneously, during any interval of observation. The immunofluorescence pattern observed in the ANA test provides a direct initial assessment of ongoing autoantibody response in candidate patients of many systemic autoimmune rheumatic diseases (SARD). As a follow-up to the International Consensus on ANA Patterns (ICAP) initiative (ANAPatterns.org), which aims to promote harmonisation of ANA pattern nomenclature and provides guidelines for ANA interpretation, thereby optimising usage in patient care, the relevance of each ANA pattern is being re-evaluated.

Methods Collective issues on ANA nomenclature were raised among research, clinical, and diagnostic laboratories represented by two workshop participants and a working committee. Post-workshop exchanges arrived at consensus on a few, but clearly not all, issues. One focus is to establish an interpretative clinical description for each defined ANA pattern for clinical use based on current literature.

Results Consensus was achieved for 28 ICAP patterns designated with alpha-numeric code (AC-1 to AC-28) and summarised under a nomenclature and classification tree categorised in three major groups (nuclear, cytoplasmic, and mitotic patterns). An important observation is that, while the Homogeneous (AC-1) and Coarse Speckled nuclear (AC-5) patterns are linked to autoantibodies strongly associated with SARD, the Dense Fine Speckled (DFS) nuclear pattern (AC-2) virtually rules out a SARD diagnosis. A clear DFS pattern is usually present when anti-DSS70/LEDGF/P75 is the only predominant autoantibody in the serum sample. DFS is the most common pattern in high titer ANA-positive, apparently healthy, individuals. Although DFS has been reported in a wide variety of chronic inflammatory diseases, such as Hashimoto’s thyroiditis, atopic dermatitis, interstitial cystitis, Vogt-Koyanagi-Harada syndrome, and in miscellaneous non-inflammatory diseases, it is not associated with SARD, even when present at very high titer.

Conclusions ICAP has clearly provided a common platform to address issues that are of great interest to the ANA community and closely linked to ANA in disease criteria. Evidently, well-defined anti-DFS ANA, confirmed by antigen-specific reflex testing, should not be considered a criterion for SLE – either in the ACR or 2012 SLICC classification criteria.