Updated antiphospholipid syndrome classification criteria

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Antiphospholipid syndrome (APS) is a systemic autoimmune disease associated with persistent antiphospholipid antibodies (aPL). It can cause thrombosis and pregnancy complications as well as non-thrombotic manifestations such as cardiac valve disease

Classification is important to help identify people to take part in research and ensures that similar disease states are included and compared in trials. This is different to tools that help clinicians make a diagnosis.

The APS classification criteria were last revised in 2006, but since then understanding of the disease has evolved. EULAR and ACR have worked together to develop a new APS classification system based on modern disease understanding. The new criteria are published in the *Annals of the Rheumatic Diseases*.

The intention was that the new tool would also allow individual criteria and different risk profiles to be weighted and demonstrate excellent operating characteristics with the highest possible specificity. Four key phases in the development of the criteria were followed: generation; item reduction; definition, further reduction, and weighting, threshold identification; and finally, validation.

After reductions and revisions, the updated classification specifies an entry criterion of at least one positive aPL test within 3 years of identification of an aPL-associated clinical criterion. This is followed by a set of weighted criteria, each scoring 1–7 points. The criteria are clustered into six clinical domains and two laboratory domains.

The clinical domains are macrovascular venous thromboembolism, macrovascular arterial thrombosis, microvascular, obstetric, cardiac valve, and hematology. The laboratory domains include assays and antibody tests to increase the specificity of the criteria. Patients scoring at least 3 points from each of the clinical and laboratory domains are classified as having APS.

The new classification criteria were tested in potential APS cases, with results showing good sensitivity and specificity. In fact, specificity for the new classification system was 99%, compared to 86% achieved with the original 2006 criteria. This is important, because classification criteria are based on standardized and stringent definitions to ensure consistency in clinical trials, and so very high specificity is required, even at the cost of sensitivity.

EULAR hopes that these new classification criteria will support high-quality, risk-stratified epidemiologic studies and clinical trials in APS, and ultimately lead to improved patient care and management recommendations.

More information: Medha Barbhaiya et al, 2023 ACR/EULAR antiphospholipid syndrome classification criteria, *Annals of the Rheumatic Diseases* (2023). DOI: 10.1136/ard-2023-224609

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