



2023 ACR/EULAR antiphospholipid syndrome classification criteria

Entry Criteria^(a)

At least one documented^(b) clinical criterion listed below (domains 1-6)

plus

A positive antiphospholipid antibody (aPL) test

(a lupus anticoagulant test, or moderate-to-high titers of anticardiolipin or anti- β_2 -glycoprotein-I antibodies [IgG or IgM]) within three years^(b) of the clinical criterion

If absent, do not attempt to classify as APS - If present, apply additive criteria

Additive clinical and laboratory criteria^(a)

Do not count a clinical criterion if there is an equally or more likely explanation than APS.
Within each domain, only count the highest weighted criterion towards the total score.

Clinical domains and criteria	Weight	Weight
D1. Macrovascular (Venous Thromboembolism [VTE]) VTE with a high-risk VTE profile ^(c) 1 VTE without a high-risk VTE profile ^(c) 3		D2. Macrovascular (Arterial Thrombosis [AT]) AT with a high-risk CVD profile ^(c) 2 AT without a high-risk CVD profile ^(c) 4
D3. Microvascular Suspected (one or more of the following) 2 Livedo racemosa (exam) Livedoid vasculopathy lesions (exam) Acute/chronic aPL-nephropathy (exam or lab) Pulmonary hemorrhage (symptoms and imaging) Established (one of more of the following) 5 Livedoid vasculopathy (pathology ^(d)) Acute/chronic aPL-nephropathy (pathology ^(d)) Pulmonary hemorrhage (BAL or pathology ^(d)) Myocardial disease (imaging or pathology) Adrenal hemorrhage (imaging or pathology)		D4. Obstetric ≥ 3 Consecutive pre-fetal (<10w) and/or early fetal (10w 0d -15w 6d) deaths 1 Fetal death (16w 0d – 33w 6d) in the absence of pre-eclampsia (PEC) with severe features or placental insufficiency (PI) with severe features 1 PEC with severe features (<34w 0d) <u>or</u> PI with severe features (<34w 0d) with/without fetal death 3 PEC with severe features (<34w 0d) <u>and</u> PI with severe features (<34w 0d) with/without fetal death 4
D5. Cardiac Valve Thickening 2 Vegetation 4		D6. Hematology Thrombocytopenia (lowest 20-130x10 ⁹ /L) 2
Laboratory (aPL) domains and criteria ^(e)		Weight
D7. aPL test by coagulation-based functional assay (lupus anticoagulant test [LAC]) Positive LAC (single – one time) 1 Positive LAC (persistent) 5		D8. aPL test by solid phase assay (anti-cardiolipin antibody [aCL] ELISA and/or anti-β_2-glycoprotein-I antibody [aβ_2GPI] ELISA [persistent]) Moderate or high positive (IgM) (aCL and/or a β_2 GPI) 1 Moderate positive (IgG) (aCL and/or a β_2 GPI) 4 High positive (IgG) (aCL <u>or</u> a β_2 GPI) 5 High positive (IgG) (aCL <u>and</u> a β_2 GPI) 7

TOTAL SCORE

Classify as Antiphospholipid Syndrome for research purposes if there are at least 3 points from clinical domains AND at least 3 points from laboratory domains