

## SUPPLEMENTARY MATERIAL

**Table S1.** Epidemiology of the Catastrophic antiphospholipid syndrome according to the literature

Diagnosis	Primary CAPS	60 %
	SLE-associated CAPS	30 %
	Others	10 %
Precipitating factors	Infections	30-50 %
	Surgery	17 %
	Malignancy	16 %
	Contraceptive	10 %
	Pregnancy-related	8-22 %
	Discontinuation of AC	unknown
	SLE flare	3 %
	Others	20 %
Mortality rate	Overall CAPS	37 %
	Primary CAPS	33 %
	SLE-associated CAPS	48 %
Laboratory features	Thrombocytopenia	67 %
	Schistocytes	22 %
	TMA <sup>a</sup>	14 %
	DIC <sup>b</sup>	11 %
	Lupus anticoagulant	83 %
	aCL IgG	81 %
	aCL IgM	49 %
	a $\beta$ 2GPI IgG	78 %
	a $\beta$ 2GPI IgM	40 %
	Antinuclear antibodies	57 %
	Anti-DNA antibodies	32 %

<sup>a</sup> TMA: Thrombotic microangiopathy defined as those patients with low platelet count, hemolysis features and schistocytes.

<sup>b</sup> DIC: Disseminated intravascular coagulation defined as those patients with low platelet count, d-dimer increase and low prothrombin time.

CAPS: catastrophic antiphospholipid syndrome; SLE: systemic lupus erythematosus; aCL: anticardiolipin antibodies; a $\beta$ 2GP1: anti-beta-2 Glycoprotein 1 antibodies