Background The epidemiology of definite antiphospholipid syndrome (APS) in the general population has not been described. A recent meta-analysis concluded that it was difficult to determine the frequency of a clinically significant antiphospholipid (aPL) profile in patients with aPL-related clinical outcomes due to the lack of robust data; only 4% of the studies had the current cutoff values for antiphospholipid antibodies (aCL) and less than one fifth of them had confirmation after 6–12 weeks. This study aimed to characterize the epidemiology of definite APS based on the 2006 updated international consensus (Sydney) classification criteria.

Methods An inception cohort of patients with incident APS in 2000–2015 in a geographically well-defined population were identified based on comprehensive individual medical record review. All cases met the definite 2006 Sydney consensus APS criteria, including the laboratory and clinical criteria as well as laboratory confirmation after 12 weeks. Lupus anticoagulant, IgM and IgG aCL and anti-2 glycoprotein-1 antibodies were tested in a centralized lab (cutoff >40 GPL/MPL). Incidence rates were age and sex adjusted to the US white 2010 population. Prevalence estimates were obtained from the incidence rates assuming no increased mortality associated with aPL and assuming migration in/out of the area was independent of disease status.

Results In 2000–2015, 33 cases of incident APS by the Sydney criteria were identified (mean age 54.2 years, 55% female; 97% Caucasian). The annual incidence of APS was 2.1 (95% confidence interval [CI]: 1.4–2.8) per 100 000 population aged 18 years. Incidence rates were similar in both sexes. The estimated prevalence of APS was 50 per 100 000 (95% CI: 42–58) and was similar in both sexes. Six (18%) patients had a concurrent diagnosis of systemic lupus erythematosus (SLE). The most frequent clinical manifestation was deep venous thrombosis. The overall mortality of patients with APS was not significantly different from the general population (standardized mortality ratio: 1.61; 95% CI: 0.74–3.05).

Conclusions Results from this first ever population based study revealed that definite APS occurred in about 2 persons per 100 000 per year. The estimated prevalence is 50 per 100 000. Overall mortality was not different from the general population. The incidence and prevalence of APS in the same population was at least as common as SLE.

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