Publication of the Month

December 12/07:   Revised APS criteria in AI patients

The antiphospholipid syndrome (APS), first described by Hughes in 1985, is a disease branching into all aspects of medicine, and linking thrombosis and autoimmunity. Since 1984 the International Congress on Antiphospholipid Antibodies has taken place every second year. Classification criteria for the antiphospholipid syndrome (APS) were formulated during the Eighth International Symposium on Antiphospholipid Antibodies in 1998 in Sapporo, Japan, and were updated in a workshop, preceding the eleventh congress in Sydney in 2004. The updated criteria were published in 2006 by Miyakis et al (see Publication of the Month March 2006). The major differences to the Sapporo-criteria are the adding of anti-ß2GPI antibodies of both the IgG and IgM isotype to the laboratory criteria and the introduction of a clear statement on threshold for positive (>40 GPL or MPL units, or >99th percentile). The interval between two serological measurements was increased from at least 6 weeks (Sapporo) to 12 weeks (Sydney).

In the following study the authors evaluated the new APS criteria with 336 samples from autoimmune patients:

Swadzba J, Iwaniec T, Szczeklik A, Musial J
Revised classification criteria for antiphospholipid syndrome and the thrombotic risk in patients with autoimmune diseases
J Thromb Haemost 2007, 5: 1883–1889

336 patients with autoimmune diseases were included in the study: 235 with systemic lupus erythematosus (SLE, according to ACR criteria), 44 with SLE-like disease (where three ACR criteria are met), 32 patients with primary APS, 12 with MCTD, 6 with systemic sclerosis, 5 with poly-/dermatomyositis, 1 with ulcerative colitis and 1 with autoimmune thrombocytopenia.
Clinical signs of APS were identified retrospectively in 180 patients (53.6%). Lupus anticoagulants (LA) were found in 95, anti-cardiolipin antibodies (aCL) in 120, and anti-ß2GPI in 101 patients (using >99th percentile). At least one antibody was found in 165 patients.

Simultaneous occurrence of both clinical and laboratory criteria for APS was present in 112 patients, including 32 with primary APS, 67 SLE patients, 11 with SLE-like syndrome and 2 with systemic sclerosis.
All antibodies examined were associated with a statistically significant risk of thrombosis. The highest risk was associated with the presence of anti-ß2GPI IgG, aCL IgG and LA (particularly "strong" LA).

The authors conclude that the updated APS classification criteria represent a step forward. However, many problems still remain and further modifications seem necessary. Currently, the main problem is the need for better identification methods of anti-phospholipid antibodies (aPL) with major clinical significance.

This month the fully automated aCL test EliA Cardiolipin will be launched – maybe one little step in the required improvement of APS diagnostics.