(107 pts) was formed of healthy blood donors and pts without presence of any signs of rheumatic or immune disease. There were 216 (89%) women and 26 (11%) men aged 10 to 80 years (mean 51.4 ± 14.6 yr.) in study group. RA group consisted of 158 (65.3%) pts, SLE – of 53 (21.9%) pts, and SS group of 31 (12.8%) pts. Diagnosis was evaluated according to standard American Rheumatology Association criteria. Enzyme immunoassay test to detect aCL IgG was performed in all pts.

Results Presence of aCL IgG was found in 38 (15.7%) pts out of 242 pts in the study group and in 6 (5.6%) control group pts. The incidence of aCL IgG was 30.2% in SLE pts, 29% in SS pts, and 8.2% in RA pts. There was significant difference in aCL IgG frequency between study pts and control groups, $p = 0.009$, and between SLE and SS pts and control pts as well ($p = 0.000014$ and 0.0002 , respectively). However, there was similar aCL IgG incidence in RA and control pts.

Age of pts with positive enzyme immunoassay (EIA) test was significantly lower as compared with negative EIA test pts (mean age 45.8 ± 15.8 yr. and 52.5 ± 14.1 yr., respectively, $p = 0.009$). However, this difference appeared to be significant in SLE pts only ($p = 0.02$).

There was no significant difference in aCL IgG incidence in separate subsets of pts either in respect to sex, or disease activity, course, or duration. In fact, some clinical manifestations were strongly associated with the presence of aCL IgG. There was significant difference in incidence of livedo reticularis, subacute vein thrombosis, and thrombocytopenia between pts with positive and negative EIA test ($p = 0.015$, 0.0066 , 0.001 , respectively).

The diagnosis of secondary antiphospholipid antibody syndrome (APS) was confirmed in 15 (6.2%) pts. There were 11 (20.7%) pts in SLE group, 2 (6.4%) pts in SS group, and 2 (1.3%) pts in RA group with this condition. The majority of those pts were aCL IgG – positive ($p = 0.0001$, as compared with aCL IgG – negative APS pts).

Conclusion 1) Presence of aCL IgG was associated with some clinical manifestations such as livedo reticularis, subacute vein thrombosis, and thrombocytopenia, and antiphospholipid antibody syndrome in pts with systemic rheumatic diseases, 2) There was no association found between presence of aCL IgG and pts sex, disease activity, course, or duration.

FRI0171 LEVELS OF SOLUBLE VCAM-1, P-SELECTIN, AND E-SELECTIN IN PATIENTS WITH PRIMARY ANTIPHOSPHOLIPID SYNDROME AND SYSTEMIC LUPUS ERYTHEMATOSUS

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Background The antiphospholipid antibodies (aPL) are known to modify the expression of adhesion molecules on activated endothelial cells and platelets *in vitro* and *in vivo*.

Objectives

Methods We studied the levels of soluble (s) P-selectin, sE-selectin, and sVCAM-1 by ELISA in sera from 19 patients (pts) (5 male, 14 female, mean age? 36.9 ± 10.1 years) with primary antiphospholipid syndrome (PAPS), 23 pts (10 male, 13 female, mean age? 35.3 ± 11.6 years) with systemic lupus (SLE) erythematosus? related antiphospholipid syndrome (SLE-APS), 15 pts

(13 male, 2 female, mean age? 29.0 ± 9.0 years) with SLE and 38 controls.

Results The sE-selectin and sVCAM-1 concentrations were significantly higher in pts with PAPS, SLE-APS and SLE compared with controls. (78.3 ± 61.2 ng/ml; 52.3 ± 14.1 ng/ml; 70.5 ± 46.9 ng/ml vs 38.0 ± 13.4 ng/ml; $p = 0.0019$, $p = 0.0363$, $p = 0.0144$, respectively for sE-selectin; 1003 ± 582.6 ng/ml; 1189.6 ± 691.0 ng/ml; 1227 ± 680.2 ng/ml vs 594.7 ± 63.3 ng/ml; $p = 0.0001$ for sVCAM-1).

The sP-selectin concentration was significantly higher in pts with SLE compared with controls (262.4 ± 158.8 ng/ml vs 120.1 ± 53.7 ng/ml; $p = 0.0037$). Within the group of pts with PAPS the sP-selectin concentration was higher among those with ischaemic CNS involvement. ($p < 0.02$).

The serum C-reactive protein level was positively correlated with the sE-selectin concentration in SLE-APS ($r = 0.341$, $p < 0.05$) and SLE ($r = 0.699$, $p < 0.01$), the sVCAM-1 concentration in PAPS ($r = 0.705$, $p < 0.001$) and SLE ($r = 0.405$, $p < 0.05$).

The serum, anti ds and ssDNA antibody level was positively correlated with the sP-selectin concentration in SLE-APS ($r = 0.730$, and $r = 0.909$, $p < 0.001$), and the sE-selectin concentration in SLE ($r = 0.684$, $p < 0.001$ and $r = 0.616$, $p < 0.002$).

Conclusion The sE-selectin and sVCAM-1 concentrations were significantly higher in pts with PAPS, SLE-APS and SLE compared with controls. (78.3 ± 61.2 ng/ml; 52.3 ± 14.1 ng/ml; 70.5 ± 46.9 ng/ml vs 38.0 ± 13.4 ng/ml; $p = 0.0019$, $p = 0.0363$, $p = 0.0144$, respectively for sE-selectin; 1003 ± 582.6 ng/ml; 1189.6 ± 691.0 ng/ml; 1227 ± 680.2 ng/ml vs 594.7 ± 63.3 ng/ml; $p = 0.0001$ for sVCAM-1).

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FRI0172 LONG-TERM FOLLOW-UP OF PAEDIATRIC PATIENTS WITH POSITIVE ANTI-PHOSPHOLIPID ANTIBODIES

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Background Long-term outcome data of Anti-Phospholipid syndrome (APLS) in paediatric patient population is very limited. There are no established criteria for making a diagnosis of Paediatric Anti- Phospholipid syndrome (APLS). Adult APLS criteria includes the major criteria of fetal loss, which is not pertinent to the pediatric population.

Objectives To evaluate the clinical outcome of paediatric patients who either had a positive Lupus Anticoagulant (LAC),

moderately high titer of anti-cardiolipin antibodies (ACLA) or anti-b₂ glycoprotein-1 antibodies on more than one occasion.

Methods We identified 19 patients (17 females, 2 males), mean age of disease onset was 11.7 years (range 7–16 years). Patients were followed-up a median of 96 months (range 12–204 months).

Results Seven patients had only IgG ACLA, one had only IgM ACLA, two had both IgG and IgM ACLA, one had IgG and IgA ACLA, and one had all three ACLA's present. One patient had significantly positive LAC and IgM ACLA and four patients had positive results for both anti- β 2 glycoprotein and ACLA. Out of 17 patients, 11 had a diagnosis of Systemic Lupus Erythematosus (SLE) and 6 patients did not have any other connective tissue disease (CTD) diagnosis. On clinical presentation, 4 patients had headaches (HA), 4 had livedo reticularis, and 8 had both HA and livedo reticularis. At initial presentation, 1 patient had venous thrombosis, 1 had chorea and one had both arterial and venous thrombosis with no previous CTD diagnosis. One patient with SLE presented with arterial thrombosis. In the follow-up period, one patient had a miscarriage and chose not to become pregnant again. All patients were placed on low-dose aspirin (ASA) [81 mg/d] and hydroxychloroquine (HCQ) if they had a diagnosis of SLE. Patients who had an event at presentation were treated with warfarin for 6 months and then switched to ASA alone after 6 months. There were no events noted in patients who were treated after their initial presentation to our clinic.

Conclusion This long-term follow-up shows that: 1) Livedo reticularis and HA are common initial presentations of APLS, but, 2) Prognosis has been very good with treatment with ASA and HCQ in the paediatric APLS group.

FRI0173 MULTIPLE ANTIPHOSPHOLIPID-ANTIBODY-TESTS IMPROVE THE LABORATORY DIAGNOSTIC IN THE ANTIPHOSPHOLIPID SYNDROME

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Background Examining patients with clinical features of the antiphospholipid-antibody-syndrome (APS), anticardiolipin antibodies (aCL) and lupus anticoagulant (LA) tests are widely performed. Unfortunately some patients with typical symptoms of APS (i.e. thromboembolic and ischaemic events, pregnancy losses) have negative test results in the commonly performed assays.

Objectives

Methods Using a new ELISA-test-system (Aesku.lab Diagnostika GmbH, Wendelsheim, Germany) we examined sera of 102 patients (68 women, 34 men) with clinical symptoms of APS: 61 patients had a history of thromboembolic events, 44 of cerebrovascular insults, 6 women had pregnancy losses. The assay detects IgG- and IgM-class-antibodies against Phosphatidyl-Serine (PS), Phosphatidyl-Inositol (PI), Sphingomyelin (SM), Phosphatidyl-Ethanolamine (PE) and Phosphatidyl-Choline (PC) without cofactor and in combination with b₂-glycoprotein I (b₂-GP-I) respectively.

Results Sera of 50 patients were completely negative in the LA- and aCL-tests, 17 patients had only low concentrations of aCL-antibodies. Of this 67 patients with negative or borderline results in the commonly performed tests, 39 (58%) were positive for

other phospholipid-antibody-specificities, in 22 (33%) the antibody-concentrations were even high. Antibodies which were tested positive in 10 or more patients are shown in the table, every "+" indicates one patient.

Abstract FRI0173 Table 1

Antibody	Strong positive sera	Weak positive sera
PS IgM without β 2-GP-I	++++++	+++
SM IgM without β 2-GP-I	++++++	++++
PE IgM without β 2-GP-I	+++	++++++
PC IgG without β 2-GP-I	++++++	++++
PC IgM without β 2-GP-I	++++++	++++

Conclusion As shown in the table, most of the phospholipid-antibody-specificities found in patients with negative or borderline results in LA- and aCL-tests are IgM-class antibodies independent from b₂-glycoprotein I. Testing for these antibodies could be helpful for the diagnostic of patients with clinical features of the APS and negative results in the commonly performed tests, especially if there are no further risk factors for thromboembolic events or ischaemic insults.

FRI0174 CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME IN A PATIENT WITH SEVERE HYPERTENSION

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Background The catastrophic antiphospholipid syndrome (CAPS) is an uncommon complication of the antiphospholipid syndrome (APS). It presents as acute multi-organ failure and more frequently involves the kidneys, lungs, gastrointestinal tract and the skin. CAPS is caused by widespread thrombosis of small and medium vessels.

Objectives

Methods Case report: We describe the case of a young female admitted with severe and accelerated hypertension. No evidence of systemic disease was observed at that time and the physical examination revealed extensive livedo reticularis. In subsequent years she also suffered several transient cerebrovascular accidents with minor neurological sequelae. The anticardiolipin antibodies (ACL) titers were usually and repeatedly extremely high. Renal angiography found normal proximal renal arteries. Renal biopsy showed ischaemic glomeruli without proliferative lesions, but with focal intimal fibrosis, thrombosis of the intrarenal vessels in the absence of vasculitis. The diagnostic of primary antiphospholipid syndrome was formulated. She was treated with antihypertensive drugs, anticoagulant and antiaggregant therapy, escalating doses of steroids, plasmapheresis.

Results Clinical course: Despite of this treatment she developed progressive multiorgan failure with malignant hypertension, renal failure, central nervous system manifestation (grand mal seizures, hemiparesis, amaurosis fugax, steady deterioration of consciousness) and gastrointestinal involvement (abdominal pain due to infarction or ischaemic ulceration of the bowel). She died as a result of a combination of renal and respiratory failure. Postmortem examination revealed extensive multiorgan arterial microthrombi.