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.

Conclusion Discussion: The case suggests that hypertension and renal failure should be recognised as a complication of APS, the aetiology being primary intrarenal vascular disease with nephroangiosclerosis. It illustrates also how primary APS presenting with renal manifestation can result in a fatal form of CAPS.

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FRI0175 CLINICAL MANIFESTATIONS OF ANTIPHOSPHOLIPID SYNDROME IN FEMALES WITH OBSTETRIC PATHOLOGY AND ANTIPHOSPHOLIPID ANTIBODIES

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Background

Objectives Antiphospholipid antibodies (APA) were studied in 94 women with a history of recurrent fetal loss and other obstetric complications associated with antiphospholipid syndrome (APS) such as fetal growth retardation, pre-eclampsia, intrauterine fetal death.

Methods

Results APA were found in 43,6% of them: lupus anticoagulant (LA) in 24,5%; anticardiolipin antibodies (aCL) ? in 10,6% and 8,5% had LA and aCL mixture. Only 4,5% of studied women had moderate or high levels of IgG aCL.

All women were examined in order to find extragenital manifestations of APS. Clinical features associated with APS including recurrent miscarriages were found in 81,6% women with APA circulation. Extragenital features of APS had 45,5% patients: recurrent arterial or venous thrombosis, thrombocytopenia in 20,4% cases. 25,1% women had possible APS features such as livedo reticularis, damage of cardiac valves, stroke, transient ischaemic attacks, migraine, leg ulcers, syndrome Reino. 29,5% APA-positive women had arterial hypertension. The most serious vascular complications took place in group with high and moderate levels of IgG aCL. One case of systemic lupus erythematosus was diagnosed (according to the American Rheumatism Association criteria).

Conclusion Women with APA circulation had not only obstetrical complications of APS. In 45,5% of them definite and possible features of this pathology were found.

FRI0176 ANTIBODIES TO CARDIOLIPIN AND B2GPI IN THE GROUP OF WOMEN'S POPULATION WITH THE HISTORY OF RECURRENT SPONTANEOUS ABORTIONS

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Background

Objectives To determine the significance of IgG anticardiolipin antibodies (aCL) and IgG antibodies to b2GPI (anti-b2GPI) in

the group of women's population with the history of recurrent spontaneous abortions.

Methods 58 women with one, two, three and more spontaneous abortions (mean age ? 29,9 ± 5,7 years) were examined. Three of them were observed out of pregnancy. In control groups 12 healthy female donors (mean age ? 29,7 ± 4,2 years) and 10 normal pregnant women (mean age? 23,2 ± 4,6 years) without history of any spontaneous abortions were included. IgG aCL and IgG anti-b2GPI were measured by ELISA.

Results The elevated levels of both autoantibodies were not noted in both control groups. In the main group in 17 (29,3%) of 58 patients (pts) IgG aCL or IgG anti-b2GPI were found. Elevated levels of IgG aCL only in 11 (64,7%) of 17 pts with antiphospholipid antibodies were noted respectively. Elevated level of IgG anti-b2GPI only wasn't noted in any pts. Elevated level of IgG anti-b2GPI and IgG aCL together had 6 (35,3%) pts.

In the group with IgG aCL without IgG anti-b2GPI (n = 11): 1) the history of one or two spont.ab. had 6 (54,5%) pts, three and more spont.ab.? 5 (45,5%) pts (mean number? 2,2 ± 1,8); 2) the clinical manifestations of the antiphospholipid syndrome (APS) were noted in 8 (72,7%), among them: 1(12,5%) pts had definite APS, 4 (50%) pts ?probable APS and 3 (37,5%) pts ? dubious APS; 3) the clinical manifestations of APS, such as deep venous thrombosis had 2 (18,2%) pts, migraine ? 7 (63,6%) pts, thrombocytopenia ? 1 (9,1%) pts. The evidences of stroke, livedo reticularis and ulcers of extremities had no one; 4) titers of antibodies were low in 3 (27,3%), medium ? in 7 (63,6%) and high ? in 1 (9,1%) pts.

In the group with IgG aCL and IgG anti-b2GPI (n = 6): 1) the history of one or two spont.ab. had 2 (33,4%) pts, three and more spont.ab.? 4 (66,7%) pts (mean number ? 5,3 ± 4,2); 2) the clinical manifestations of APS were noted in 6 (100%) pts, among them: 4 (66,7%) had definite APS, 2 (33,3%)? probable APS and no one had dubious APS; 3) the clinical manifestations of APS, such as thrombosis (arterial and venous, deep and superior) had 2 (33,3%) pts, migraine?5 (83,3%), thrombocytopenia? 5 (83,3%), stroke ? 2 (33,3%), livedo reticularis? 4 (66,7%) and ulcers of extremities? 1 (16,7%) pts; 4) titers of both antibodies were medium and high.

Conclusion We found that IgG aCL and IgG anti-b2GPI, especially their combination is one of the most important tests for APS in the pregnant women's population with the history of spontaneous abortions, especially among the women with habitual abortion; simultaneous production of both antibodies correlated with severity of clinical manifestations of APS. The discovery of IgG anti-b2GPI make the clinical manifestations of APS more severe.

FRI0177 THE SPECTRUM OF CLINICAL MANIFESTATIONS OF ANTIPHOSPHOLIPID ANTIBODY SYNDROME IN THE GENERAL HOSPITAL

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Background APS is recognised with growing incidence in many fields of medicine. It is clinically and immunologically diverse condition and may affect any organ of the body and display a broad spectrum of manifestations. The majority of published studies have been reported from a tertiary referral hospitals. We