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Antiphospholipid syndrome

What is antiphospholipid syndrome?

Antiphospholipid syndrome (APS) is a disorder characterised by recurrent arterial or venous thrombosis and/or pregnancy losses, in the presence of persistently elevated levels of anticardiolipin antibodies and/or evidence of circulating lupus anticoagulant (these abnormalities are detected by blood tests).

Primary APS occurs when there is no evidence of associated diseases. APS in the presence of an underlying disease, usually systemic [lupus erythematosus](#), is called secondary APS.

Who gets APS?

APS occurs more commonly in young to middle-aged adults. However, it has also been found in a child as young as 8 months and in elderly people. Secondary APS appears to be more common in females and is most likely due to the fact that systemic lupus erythematosus and other connective tissue disorders have a female predominance.

Younger patients with a history of deep vein thrombosis (DVT), pulmonary embolism, myocardial infarction, or cerebrovascular accidents (CVA), need to be investigated for APS, particularly if no other risk factors are present.

What are the signs and symptoms?

Signs and symptoms of APS are many and varied but to classify as APS a patient must have at least one of the two following clinical manifestations in addition to the presence of certain laboratory abnormalities.

- Venous or arterial thrombosis: this may involve the cerebral vascular system, coronary arteries, pulmonary emboli or thromboses, hepatic or renal veins, ocular veins or arteries
- Recurring miscarriages or premature births: patients may have pre-eclampsia in pregnancy and babies may be unexpectedly small

Antiphospholipid syndrome



Livedo



Splinter haemorrhages



Vasculitis



Other symptoms, although not part of the classification criteria, include:

- Skin disorders: [livedo reticularis](#) (seen in up to 80% of people with antiphospholipid antibodies), splinter haemorrhages (red or black streaks found on nails), leg ulcers, superficial thrombophlebitis, blue toe syndrome, [vasculitis](#)
- Neurological defects: migraine headaches, seizures, multi-infarct dementia
- Cardiac abnormalities: heart murmur, cardiac valve vegetations
- Blood abnormalities: thrombocytopenia (low platelet count), haemolytic anaemia (low red cell count due to destruction of the cells by antibodies)

A severe form of the disease is called catastrophic antiphospholipid syndrome. This is where blockage of blood vessels in multiple organs may occur over days or weeks. The condition is serious and often lethal.

Diagnosis of APS

Diagnosis of APS is confirmed from laboratory tests that show the presence of antiphospholipid antibodies or abnormalities in phospholipid-dependent tests of blood clotting (coagulation). Patients with suspected APS should be tested for the following:

- aCL antibodies
- Anti-beta-2 glycoprotein I antibodies
- Activated partial thromboplastin time (aPTT)
- LA tests such as dilute Russell viper venom time (DRVVT)
- False-positive serologic test result for syphilis
- Complete blood cell count (thrombocytopenia, Coombs-positive haemolytic anaemia)

What treatment is available?

The main aim of treatment is to prevent the clinical manifestations of APS. Risk factors for thrombosis should be identified and removed or corrected, for example, smoking, oral contraceptives, high blood pressure or elevated blood fats. Antiplatelet drugs such as low dose aspirin may be useful.

There is no specific treatment for APS. Signs and symptoms are treated as they occur. For example, a thrombosis or embolism is treated with the anticoagulants heparin and warfarin.

Related information

References:

On DermNet NZ:

- [Livedo reticularis](#)
- [Lupus erythematosus](#)

Other websites:

- [APS Foundation of America, Inc.](#)
- [Antiphospholipid Syndrome](#) - emedicine, the online textbook

Books:

See the [DermNet NZ bookstore](#)

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DermNet does not provide an on-line consultation service.
If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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