Antiphospholipid Syndrome

Antiphospholipid syndrome (APLS, APS) is an autoimmune condition thought to be caused by a heterogeneous group of antibodies directed against various phospholipid-associated proteins, such as those found in the cell membrane. The presence of these antibodies leads to hypercoagulability, which may manifest as recurrent thromboembolisms and miscarriages. Definitive diagnosis requires the identification of the antibodies in question. These may be anticardiolipin antibodies, anti-beta-2-glycoprotein antibodies, or lupus anticoagulant. Management is achieved with anticoagulation.



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Characteristics

Antiphospholipid Antibodies

Ant-tie-body Pointing to Phospholipid Door

Antiphospholipid syndrome is thought to be caused by a heterogenous group of antibodies directed against various proteins associated with phospholipids, such as those found in the cell membrane. The action of these antibodies then leads to activation of the clotting cascade.

Hypercoagulability

Hiker-clogs

The presence of antiphospholipid antibodies in the blood often leads to hypercoagulability. This effect is multifactorial, but is due in part to increased tissue factor expression, increased oxidative stress, activation of complement, and inhibition of fibrinolysis.

Clinical Features

Recurrent Thromboembolisms

Recurrent-clock Trombone-Elmo

Antiphospholipid Syndrome manifests clinically as recurrent thromboembolisms, which can occur in the venous circulation in the form of deep vein thromboses (DVT) and pulmonary embolism (PE), or arterial circulation as in transient ischemic attack (TIA)/stroke, and myocardial infarction (MI).

Recurrent Miscarriages

Recurring Missed-carriage

One of the common clinical manifestations associated with Antiphospholipid Syndrome is recurrent miscarriage. The underlying pathophysiology is not completely understood, however it is thought that the activation of coagulation caused by antiphospholipid antibodies may play a role by leading to poor circulation to the placenta.

Diagnosis

Anticardiolipin Antibodies

Cardio-heart-lips Ant-tie-body

Diagnosis of Antiphospholipid Syndrome consists of testing for the presence of certain antibodies, of which there are several specifically associated with Antiphospholipid Syndrome, including anticardiolipin antibody.

Anti-Beta-2-Glycoprotein Antibodies

Beta-fish (2) Tutu Glider-protein Ant-tie-bodies

Anti-Beta-2-Glycoprotein antibodies are antibodies that bind to the glycoproteins and phospholipids found in the cell membranes. Binding of Anti-Beta-2-Glycoprotein antibodies to the cell membranes initiates the clotting cascade and predisposes the patient to thrombosis.

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Lupus Anticoagulant

Ant-tie-clogs watching Lupus-loop butterfly

Diagnosis of Antiphospholipid Syndrome consists of testing for the presence of certain antibodies, of which there are several specifically associated with Antiphospholipid Syndrome, including Lupus Anticoagulant. Importantly, the term "Lupus Anticoagulant" is actually a misnomer and refers to the fact that it has an anticoagulant effect during in-vitro testing in labs, however in the body it has a pro-coagulant effect.

Management

Anticoagulation

Ant-tie-clogs with Treats

In patients found to have an antibody titer consistent with antiphospholipid syndrome and who have had clinical manifestations such as recurrent thrombosis or miscarriages, long-term anticoagulation with Warfarin is indicated. For these patients, Warfarin is preferred over direct oral anticoagulants (DOACs) as it has been shown to be more effective, though the exact reason why is not well known. Patients with acute thromboembolism due to antiphospholipid syndrome may be managed with heparin bridged to warfarin.

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