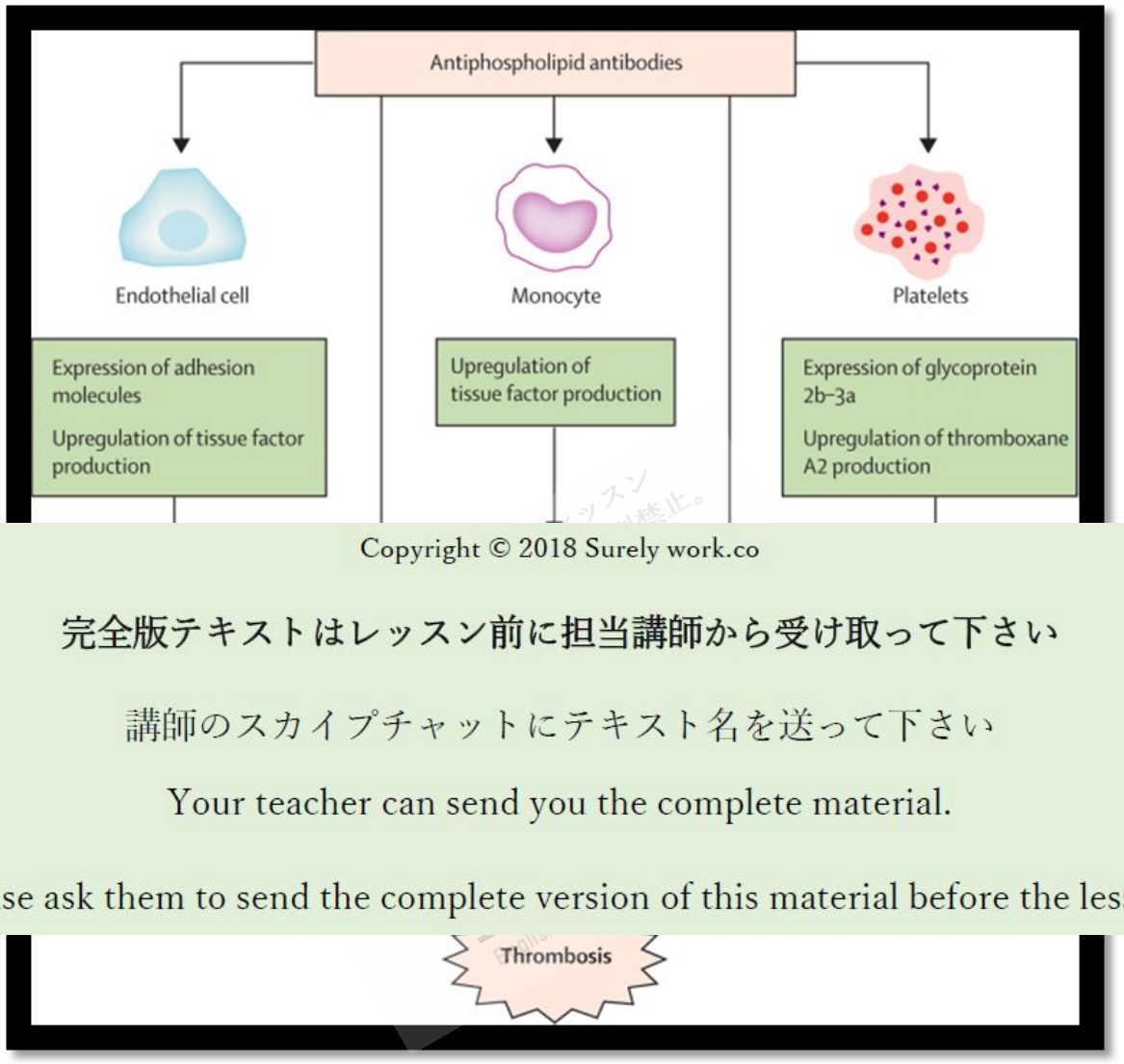
Antiphospholipid Syndrome



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What is antiphospholipid syndrome?

The antiphospholipid syndrome is a disorder of the immune system that is characterized by excessive clotting of blood and/or certain complications ofpregnancy (premature miscarriages, unexplained fetal death, or premature birth) and the presence of antiphospholipid antibodies (cardiolipin or lupusanticoagulant antibodies) in the blood. Patients with antiphospholipid syndrome have developed abnormal symptoms while having antiphospholipid antibodies that are detectable with blood testing.

Antiphospholipid syndrome is also called phospholipid antibody syndrome. Antiphospholipid syndrome has been referred to as Hughes syndrome in honor of the doctor who first described it.

It is important to note that antiphospholipid antibodies can also be found in the blood of individuals without any disease process. In fact, antiphospholipid antibodies have been reported in approximately 2% of the normal population. Harmless antiphospholipid antibodies can be detected in the blood for a brief period occasionally in association with a wide variety of conditions, including bacterial, viral (hepatitis, HIV), and parasite (malaria) infections. Certain drugs can cause antiphospholipid antibodies to be produced in the blood, including antibiotics, cocaine, hydralazine,procainamide, and quinine.

Nevertheless, the antiphospholipid antibody (a protein) is not considered a normal blood protein and has been found in patients to be associated with a number of illnesses. These illnesses include abnormal clotting (thrombosis) of arteries (stroke, infarction) and/or veins (phlebitis), premature miscarriages (spontaneous abortions), abnormally low blood platelet

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Patients with the antiphospholipid syndrome can have a variety of antibodies to molecules called phospholipids in their blood. These antibodies include VDRL/RPR (a syphilis test that can be falsely positive in these patients), lupus anticoagulant, prolonged PTT, and cardiolipin antibody. As mentioned above, the anticardiolipin antibody has also been found in patients with the immune disease systemic lupus erythematosus, which is characterized by the production of a variety of abnormal antibodies.

What causes antiphospholipid syndrome?

The cause of antiphospholipid syndrome is not completely known. Antiphospholipid antibodies reduce the levels of annexin V, a protein that binds phospholipids and has potent clot-blocking (anticoagulant) activity. The reduction of annexin V levels is thought to be a possible mechanism underlying the increased tendency of blood to clot and the propensity to pregnancy loss characteristic of the antiphospholipid syndrome.

Antiphospholipid antibodies, such as anticardiolipin, have also been associated with decreased levels of prostacyclin, a chemical that prevents the clumping together of normal blood clotting elements called platelets.

How is antiphospholipid syndrome treated?

The treatment of patients with anticardiolipin syndrome has substantially evolved since they were discovered to be clinically important in the mid-1980s. Each manifestation of the antiphospholipid syndrome, and each individual patient with the condition, is treated uniquely.

Because many of the features of illness with anticardiolipin syndrome are associated with an abnormal grouping of normal blood clotting elements (platelets), treatment is often directed toward preventing clotting by thinning the blood. Patients with this disorder have an abnormal tendency to formblood clots (thrombosis). The abnormal blood clotting can affect the function of virtually any organ. Medications that thin (anticoagulate) the blood, such as heparin (Hep-Lock, Liquaemin) and warfarin (Coumadin) (powerful blood thinners), are used for treatment. Aspirin has an affect on platelets that inhibits their grouping (aggregation) and has also been used in low doses to thin the blood of selected patients. Cortisone-related medications, such asprednisone, have been used to suppress the immune activity and inflammation in patients with certain features of the condition. For patients with systemic lupus erythematosus who also have antiphospholipid syndrome, hydroxychloroquine (Plaquenil) has been reported to add some protection against blood clotting.

Other reported treatments include the use of intravenous gamma globulin for selected patients with histories of premature miscarriage and those with low blood-clotting elements (platelets) during pregnancy. Recent research studies, however, suggest that intravenous gamma globulin may be no more effective than combination aspirin and heparin treatment.

What is catastrophic antiphospholipid syndrome?

Catastrophic antiphospholipid syndrome is a variant of antiphospholipid syndrome that is characterized by blockage of many blood vessels throughout the body. As a result of catastrophic antiphospholipid syndrome, many organs can be affected, including the skin, lungs, brain, heart, kidneys, and bowels. Catastrophic antiphospholipid syndrome is treated with anticoagulation, corticosteroids (cortisone medication), andplasmapheresis (plasma exchange).

Catastrophic antiphospholipid syndrome is rare, affecting less than 1% of those with antiphospholipid syndrome. Catastrophic antiphospholipid syndrome is sometimes referred to as Asherson's syndrome after the researcher who described it in the early 1990s.

Antiphospholipid Syndrome at a Glance

- Antiphospholipid syndrome is an immune disorder that can affect virtually any organ.

- Patients with antiphospholipid syndrome can have a variety of antibodies to phospholipids in their blood.
- Antiphospholipid syndrome involves abnormal tendency toward clotting of blood.
- Each individual patient with the antiphospholipid syndrome is treated uniquely according to what symptoms are present.

Reference:
http://www.medicinenet.com

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