CONDITION

Antiphospholipid syndrome - APS

Learn how antiphospholipid syndrome can cause blood clots and pregnancy complications, how APS is diagnosed, and how treatment lowers future risks.

WHAT IT IS
Autoimmune blood-clotting condition

BODY SYSTEM
Blood and circulation

MAIN SYMPTOMS
One-sided leg swelling, leg pain, shortness of breath, chest pain

WHAT CAUSES IT
Immune system reaction

HOW IT IS DIAGNOSED
Clinical event and repeat antibody blood tests

MAIN TREATMENT
Anticoagulant medicines

CAN IT BE CURED?
No, treatment lowers future risks

IS IT AN EMERGENCY?
Sometimes, with sudden shortness of breath

LONG-TERM OUTLOOK
Treatment reduces risks but recurrent clots can occur

Overview

Antiphospholipid syndrome (APS) is an autoimmune condition that increases the risk of abnormal blood clots and certain pregnancy complications. In an autoimmune condition, the immune system mistakenly reacts against parts of the body.

People with APS have specific proteins in their blood called antiphospholipid antibodies. These antibodies react with proteins that bind to phospholipids, which are normal components of cells. Their presence can affect clotting, blood vessel walls, and the placenta.

APS may cause a clot in a vein, such as a deep vein thrombosis in the leg, or in an artery, such as an artery supplying the brain. During pregnancy, APS can interfere with placental function and increase the risk of miscarriage, fetal loss, preeclampsia, poor fetal growth, or early delivery.

Some people have antiphospholipid antibodies without ever having a clot or pregnancy complication. A positive blood test alone does not necessarily mean that a person has APS.

APS can occur by itself. This is sometimes called primary APS. It can also occur with another autoimmune disease, especially systemic lupus erythematosus, often called lupus. Treatment can substantially reduce the risk of further clots and improve pregnancy outcomes.

Symptoms

APS does not usually cause symptoms unless it leads to a clot, pregnancy complication, or another effect on blood vessels or blood cells. Symptoms depend on which part of the body is affected.

A deep vein thrombosis, or DVT, most often develops in a leg. Possible symptoms include:

Contact a healthcare professional urgently for possible DVT symptoms. Prompt assessment is important because part of a clot can travel to the lungs.

A pulmonary embolism is a clot blocking blood flow in the lungs. Call emergency services for sudden shortness of breath, chest pain, coughing up blood, fainting, or a very fast heartbeat.

A clot affecting the brain can cause a stroke or transient ischemic attack, sometimes called a mini-stroke. Call emergency services immediately for facial drooping, weakness or numbness on one side, difficulty speaking, sudden loss of vision, severe loss of balance, or a sudden unusual headache.

Pregnancy-related APS may be identified after repeated pregnancy losses or complications linked to placental problems. These can include fetal growth restriction, preeclampsia, or medically necessary early delivery. A person may not have obvious symptoms before a pregnancy complication occurs.

Other findings associated with APS can include:

During pregnancy, seek urgent obstetric advice for vaginal bleeding, reduced fetal movement, a severe or persistent headache, vision changes, sudden swelling, severe upper abdominal pain, chest pain, or breathing difficulty.

- New swelling in one leg
- Leg pain or tenderness
- Warm or discolored skin over the affected area
- A lace-like, purple or reddish skin pattern called livedo reticularis
- Low platelets, which are blood cells involved in clotting

- Heart valve abnormalities
- Kidney problems caused by injury to small blood vessels
- Headaches or migraine, although these are common and do not by themselves establish APS

Causes and risk factors

The exact reason APS develops is not fully understood. It results from an interaction between the immune system, blood clotting pathways, and blood vessel cells. APS is not contagious.

The three main types of antiphospholipid antibodies used in assessment are lupus anticoagulant like warfarin or apixaban , anticardiolipin antibodies, and anti-beta-2 glycoprotein I antibodies. Despite its name, lupus anticoagulant is not an anticoagulant medicine and does not mean a person has lupus. In the body, it is associated with an increased clotting risk.

Some antibody patterns carry more risk than others. For example, persistently positive results for all three antibody groups are often described as "triple positivity" and are associated with a higher clot risk. A specialist interprets the pattern together with the person's medical history.

APS may occur without another known condition, or it may be associated with lupus or another autoimmune disease. Family members may share a tendency toward autoimmune disease, but APS does not follow a simple pattern of inheritance.

In people with antiphospholipid antibodies, other situations can add to clot risk. These include major surgery, a hospital stay, prolonged immobility, pregnancy, the weeks after delivery, smoking, estrogen-containing medicines, and uncontrolled cardiovascular risk factors. Infection and inflammation may also contribute.

Antiphospholipid antibodies can sometimes appear temporarily during an infection or in other situations. This is one reason that repeat testing is needed before persistent antibodies are confirmed.

Diagnosis

There is no single test that diagnoses APS on its own. Doctors consider both the person's medical history and laboratory results.

Assessment usually begins with a detailed history of blood clots, strokes, pregnancy complications, autoimmune symptoms, medicines, and other clotting risks. Medical records or imaging reports may be reviewed to confirm previous events.

Blood tests look for:

To support an APS diagnosis, an antiphospholipid antibody generally needs to remain positive on tests performed at least 12 weeks apart. Repeating the test helps distinguish persistent antibodies from a temporary result.

Testing can be difficult to interpret. Anticoagulant medicines, acute illness, and laboratory methods may affect some results, especially lupus anticoagulant testing. A hematologist or another clinician experienced in APS may help plan and interpret testing. People should not stop an anticoagulant for testing unless the clinician managing it provides specific instructions.

APS is generally diagnosed when a person has an appropriate clinical event, such as a confirmed clot or certain pregnancy complications, together with persistent qualifying antibodies. Classification criteria developed for research can guide evaluation, but they are not a substitute for clinical judgment.

Imaging may be needed to confirm a suspected clot. Depending on the symptoms, this might include an ultrasound of a leg, a scan of the lungs, or brain imaging. Other tests may evaluate blood counts, kidney function, urine, heart valves, or possible autoimmune disease.

- Lupus anticoagulant
- Anticardiolipin IgG and IgM antibodies
- Anti-beta-2 glycoprotein I IgG and IgM antibodies

Treatment and management

Treatment depends on whether a person has had a venous clot, an arterial clot, pregnancy complications, or only positive antibodies. Other medical conditions and bleeding risks also matter.

A new blood clot requires prompt medical treatment. Initial treatment often uses a fast-acting anticoagulant, commonly called a blood thinner. These medicines do not dissolve a clot directly. They prevent it from growing while the body gradually breaks it down.

For thrombotic APS, meaning APS that has caused a clot, long-term treatment commonly involves a vitamin K antagonist such as warfarin. Regular blood tests are needed to keep its effect within the target range chosen by the treating team. The length and intensity of treatment depend on the type of clot, whether it was triggered by a temporary risk, the antibody profile, recurrence history, and bleeding risk.

Direct oral anticoagulants are used for many other clotting conditions, but they are not generally preferred for people with high-risk APS, especially those with triple-positive

antibodies or a previous arterial clot. Treatment decisions should be made with a clinician familiar with APS. Do not change or stop anticoagulant treatment without medical advice.

Management after an arterial event, such as a stroke, may involve a vitamin K antagonist and, in selected cases, an antiplatelet medicine such as aspirin. The balance between preventing clots and avoiding bleeding must be considered individually.

A person with positive antibodies but no previous clot or qualifying pregnancy complication does not automatically need anticoagulation. Some people with higher-risk antibody patterns may be advised to take low-dose aspirin, particularly when other risks are present, but this decision requires clinical assessment.

For pregnancy-related APS, treatment often includes low-dose aspirin and an injectable heparin medicine. The exact plan depends on whether there has been a previous clot and on the pregnancy history. Heparins do not cross the placenta. Warfarin is generally avoided during pregnancy because it can harm the developing fetus, although specialist exceptions may apply in complex circumstances.

Pregnancy should ideally be planned with an obstetric team and the clinician managing APS. Monitoring may include blood pressure checks, blood tests, and ultrasound assessment of fetal growth and placental function. Anticoagulant planning is also needed for delivery and the postpartum period, when clot risk remains increased.

Everyday management includes addressing other clot risks. This may involve stopping smoking, staying active when medically safe, maintaining a healthy blood pressure and cholesterol level, and avoiding long periods without movement. Before starting estrogen-containing contraception or menopausal hormone therapy, discuss the clot risk and alternatives with a clinician.

Anticoagulants can cause bleeding. Patients should tell healthcare professionals and dentists that they take an anticoagulant. New medicines, supplements, illness, and major dietary changes may affect warfarin. Unusual bleeding, black stools, vomiting blood, a major head injury, or bleeding that will not stop requires urgent medical assessment.

Complications

Without effective prevention, APS can lead to recurrent clots. Possible long-term effects depend on the location and severity of the event.

A leg DVT can damage veins and cause ongoing swelling, discomfort, or skin changes, known as post-thrombotic syndrome. A pulmonary embolism may occasionally lead to persistent high pressure in the lung arteries. A stroke can affect movement, speech, memory, vision, or

independence.

Placental complications can result in pregnancy loss, impaired fetal growth, preeclampsia, or premature delivery. Specialist treatment and monitoring can improve outcomes, although they cannot remove every risk.

Catastrophic APS is a rare, severe form in which clots rapidly affect several organs, often over days. Symptoms vary but can include severe breathing problems, confusion, kidney problems, abdominal symptoms, or widespread skin changes. It is a medical emergency requiring hospital treatment. Management may combine anticoagulation with treatments that reduce immune activity or remove harmful antibodies from the blood.

Follow up

Routine follow-up helps the care team review clot prevention, bleeding risk, pregnancy plans, cardiovascular health, and any associated autoimmune disease. Care may involve primary care, hematology, rheumatology, neurology, or a high-risk pregnancy team.

People taking warfarin need regular clotting tests and medication reviews. Monitoring may be more frequent after a dose change, illness, or the start of another medicine. A later negative antibody test does not automatically mean treatment can be stopped.

Contact the treating team before surgery, dental procedures, long-distance travel, pregnancy, or starting hormonal medication. Anticoagulant treatment may need advance planning, but it should not be interrupted without instructions.

Routine appointments are appropriate for medication questions, mild side effects, pregnancy planning, or discussion of future clot risks. New one-sided leg swelling needs urgent assessment. Stroke symptoms, severe breathing difficulty, significant chest pain, coughing up blood, fainting, or uncontrolled bleeding require emergency care.

Sources

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