

Antiphospholipid Antibody Syndrome



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~ Introduction ~

This pamphlet is a layman's terms summary of Antiphospholipid Antibody Syndrome (APS). It covers such topics as diagnosis, symptoms, treatment, and coping. It is meant for patients newly diagnosed, however, would also be good for informing friends and family about your disease.

~ What is Antiphospholipid Antibody Syndrome ~

Antiphospholipid Antibody Syndrome or APS is an autoimmune disorder in which the body recognizes certain normal components of blood and/or cell membranes as foreign substances and produces antibodies against them. Patients with these antibodies may experience blood clots, including heart attacks and strokes, and miscarriages. APS may occur in people with systemic lupus erythematosus, other autoimmune diseases, or in otherwise healthy individuals.

APS is also known as APLS, APLA, Hughes Syndrome or "Sticky Blood".

~ APS is an autoimmune disease ~

One way in which our immune system fights infections is by making antibodies. Antibodies are proteins in the blood and body fluids that bind to foreign invaders like bacteria and viruses and help the immune system destroy and remove them. Sometimes the immune system doesn't function properly and makes antibodies against normal organs and tissues in the body. These self-reactive antibodies are called autoantibodies. The autoantibodies in APS were originally thought to recognize that recognize certain phospholipids, fatty molecules that make up part of normal cell membranes, hence the name "antiphospholipid" antibodies. It is now known that most of the autoantibodies in APS patients actually recognize certain blood proteins that bind to phospholipids, not the phospholipids themselves. Two blood proteins that are major targets of antiphospholipid antibodies are b₂-glycoprotein I and prothrombin.

~ What is CAPS? ~

Catastrophic antiphospholipid antibody syndrome is a very rare complication encountered in a subset of patients with antiphospholipid antibody syndrome. This rare syndrome is characterized by the development of multiple blood clots that block small blood vessels in several organs in the body. The organs most commonly affected by these small blood clots include the heart, lungs, nervous system, and kidneys. In many ways, this syndrome is similar to another rare disease, thrombotic thrombocytopenic purpura.

~ How is APS Diagnosed ~

Physicians use a combination of clinical symptoms (see above) and laboratory tests to diagnose APS. The common blood tests for antiphospholipid antibodies are as follows:

Anticardiolipin antibodies (IgG, IgM, and IgA)

Lupus anticoagulant – a panel of blood clotting tests that may include the dilute Russel Viper venom time (dRVVT), lupus aPTT, mixing studies, and hex phase phospholipid test, platelet neutralization procedure

Antibodies to b₂-glycoprotein I (IgG, IgM, IgA)

Panels of tests for antibodies to phospholipids other than cardiolipin are available but have not undergone the rigorous international standardization efforts applied to anticardiolipin assays. A number of experts in the field question the usefulness of these panels, which may be quite expensive.

~ Clinical Features of APS ~

People with antiphospholipid antibodies have an increased risk of developing one or more of the following problems:

Blood clots in veins, particularly deep vein thrombosis (DVT)

Blood clots that go to the lungs (pulmonary embolism)

Blood clots in arteries

Miscarriages – these can occur at any stage of pregnancy but are most common in the late first trimester or early second trimester

Pre-eclampsia, eclampsia, fetal growth retardation, premature delivery

Heart attacks, angina

Strokes

Brief stroke-like episodes called transient ischemic attacks (TIAs), for example, loss of vision

Decreased levels of platelets (small blood cells involved in blood clotting)

Heart valve problems, sometimes requiring valve surgery or valve replacement

Persistent or transient blotchy, lacy bluish rash (called livedo reticularis)

Skin ulcers, most commonly on the legs or feet

“Catastrophic” APS – a very rare, life-threatening syndrome in which clots form in small blood vessels of multiple organs (such as heart, lungs, brain, kidneys)

Other complications/symptoms that *might* be associated with antiphospholipid antibodies include:

Problems with thinking clearly (loss of concentration, difficulty with reading comprehension and performing calculations, memory loss)

Neurological problems similar to multiple sclerosis

Migraine headaches, sometimes with visual disturbances

Other neurological symptoms including episodes of partial or total vision loss, dizziness, vertigo, loss of balance, seizures, and other abnormal movements

Start a medical journal to keep track of any events. List the date, time, symptom, how long it lasted, severity, and anything you think that might have triggered it. For rashes, and color changes start a photo journal. This is always helpful to show to the doctors, since the rash or color change may be gone by the time you have an appointment to see one. Write down any questions you may have and take the journal with you to your appointments.

~ APS Treatment ~

There is no cure for APS, but there is treatment. The treatment of choice for patients with APS who have had a blood clot is anticoagulant therapy. This is usually successful in preventing further clots. For women with APS and recurrent miscarriages who have not had a prior blood clot, the use of anticoagulant therapy during the pregnancy significantly increases the likelihood of a successful outcome.

Some individuals may have elevated antiphospholipid antibodies but have no clinical manifestations of the syndrome. These individuals are usually treated with aspirin. Aspirin reduces the risk of blood clots by making the platelets less sticky. Studies are ongoing to determine how helpful aspirin is and whether low doses of anticoagulants might be more effective.

In general patients who have had a blood clot (i.e., stroke, heart attack, DVT) and have persistently positive tests for antiphospholipid antibodies should be treated with anticoagulants indefinitely. Discontinuing treatment after a fixed period of time, such as six months, may be quite dangerous in such patients. In some patients with a history of blood clots, antiphospholipid antibodies may disappear after a certain period of time. It is not known whether it is safe to stop anticoagulation in this situation. Consultation with a doctor experienced in treating APS is recommended for such patients.

~ Doctors and Information on APS ~

Although APS is actually one of the more common autoimmune diseases, some primary care doctors remain uninformed about it. When their patients have symptoms of APS, these doctors may not test for antiphospholipid antibodies soon enough or at all. Unfortunately, many patients have had to see several physicians and specialists before getting the proper diagnosis and treatment.

The type of doctor a patient sees should be determined by the symptoms the individual patient is having, and any given patient may benefit from the input of several specialists. For patients with blood clots, a hematologist would be involved, often for management of anticoagulant therapy (blood thinner). For patients with recurrent miscarriages, a high-risk obstetrician should be consulted. For patients who also have rheumatologic symptoms, such as symptoms of lupus, a rheumatologist would be important to see. Of course, all patients would benefit from having a single physician identified as their primary care provider, to help coordinate all of their healthcare needs.

~ APS: The Statistics ~

15-20% of all cases of blood clots in large veins (deep vein thrombosis) including blood clots that go to the lungs (pulmonary embolism) are due to APS

10-25% of women with recurrent miscarriages have APS

33% of strokes occurring in younger people (under the age of 50) are due to APS

APS is a major women's health issue: 75-90% of those affected by APS are women

40-50% of patients with lupus also have APS

1-5% of the general population is believed to have APS

APS and Pregnancy Complications

Women with APS may have difficulties with pregnancy. During pregnancy, women are at higher risk of developing blood clots and preeclampsia. In APS, pregnancies are thought to be lost because blood clots form in the placenta and starve the baby of nutrition. Some women may have trouble getting pregnant, while others may experience repeated miscarriages. Blood clots that develop in the placenta can cause fetal growth problems, fetal distress, pre-term birth, or pregnancy loss.

Expert care and close monitoring of the pregnancy is essential by a doctor knowledgeable about APS. During pregnancy, physicians may recommend low doses of aspirin and daily injections of the blood thinning drug, heparin. This gives the fetus about an 80% chance of survival, a drastic improvement from the 1980s when fetal survival was around 20%. The therapy is started at the beginning of pregnancy and halted just before delivery to reduce the risk of bleeding during childbirth. Soon after birth, the treatment resumes for about six weeks because of an increased risk for clotting in the postpartum period. In a more serious case, preeclampsia may set in towards the end of pregnancy, and a planned premature birth may be necessary. Heparin can cause bone loss, so women may need to take additional calcium during pregnancy. In addition, women need to be monitored for development of a low platelet count.

Over the long term, many doctors recommend women continue to take a low dose of aspirin to reduce the risk of developing dangerous blood clots. Many women with APS are unaware they have the condition, but it can be diagnosed with a blood test. Doctors may consider the diagnosis when a woman has repeated, unexplained pregnancy loss.

If you are trying to get pregnant or are pregnant, it is very important to let your doctor know immediately. Continued use of warfarin may cause birth defects. The doctor will change your medication to a different blood thinner that is safe. Using proper treatment, women with APS have about the same risks as other women during pregnancy.

APS pregnancies are not normal. Normal pregnancy is 40 weeks. In APS, it is more common to deliver the baby between 30-35 weeks, and between 3-5 pounds. Heparin protects the placenta partially, but not fully, so that the baby gets enough nutrition to survive longer in the mother. Once born, the babies do fine.

Many women who have problems with APS during pregnancy are completely fine when not pregnant. Others do go on to develop problems with clotting. Currently there is no way of telling which women will be unlucky, until a clot actually occurs.

Infertility has also been linked to antiphospholipid antibodies. Testing for these antibodies is becoming routine in infertility clinics.

Birth Control Pills & Hormone Replacement Therapy

Women also need to avoid estrogen therapy (such as birth control or hormone replacement therapy) because estrogen predisposes patients to clotting.

Other forms of contraception should be discussed with your doctor.

Problems with Periods

Some women taking warfarin experience problems with increased bleeding. It can lead to anemia. Tell your doctor about this problem. The doctor can recommend several options and prevent anemia. One example is: for women who have already given birth and are not actively trying to conceive, the Mirena® IUD has been successful in reducing period blood loss. As it only releases hormones to the uterus lining and is not absorbed into the blood stream, therefore, it is safe for women with APS to use.

~ Other Points to Consider ~

You may notice you bruise more easily or little cuts will bleed longer when you are taking warfarin (Coumadin®). Injuries can be more serious when on anticoagulants and care should be taken during any activity that can result in injury. Contact sports are not recommended.

If, while on anticoagulants, you injure your head, go directly to the Emergency Room. Your brain is very sensitive to bleeding while on anticoagulants.

If a serious injury does occur go directly to the Emergency Room and be sure they know you are taking anticoagulants and tell them what your most recent INR was.

You should have a medical alert bracelet and wear it at all times. You can order one through Medic Alert® at <http://www.medicalert.org>. A bracelet is the most visible, the easiest and the most recognized however, there are also necklaces and other types available.

~ Safety When Traveling ~

Long trips, especially by air, have some clotting risk even for non-APS people. It is important for people with APS to get up and walk around at least every couple of hours. On long car trips stop at least every two hours and walk. Drink plenty of water and wear compression stockings to help reduce your chance of DVT. If you plan to be away during the time of a periodic blood test, arrange for the blood test before you leave for the trip.

~ Take Your Medication ~

It is very important to take your medicine every day. Try to take your the medicine at the same time each day for consistency. You may want to get a pillbox that holds at least one week's supply of the pill(s) or to mark it on a calendar when you take your medicine. This will help you to know when you have taken your pills. Do not take two doses in one day if you have forgotten your dose.

~ Symptoms to Watch For ~

If you test positive for APS antibodies you should be aware of the symptoms caused by blood clots. If any of these symptoms occur, seek medical help immediately. Symptoms that could be caused by a blood clot include:

Heart Attack: Chest discomfort or pain. Most heart attacks involve discomfort in the center of the chest that lasts more than a few minutes, or that goes away and comes back. It can feel like uncomfortable pressure, squeezing, fullness, or pain. The pain generally becomes so bad that it may feel unbearable and relentless, but occasionally the pain can be milder.

Discomfort in other areas of the upper body. Symptoms can include pain or discomfort in one or both arms, the back, neck, jaw, or stomach

Shortness of breath. This feeling often comes along with chest discomfort. But it can occur before the chest discomfort

Other signs may include breaking out in a cold sweat, nausea, or light-headedness

Stroke: (Blood Clot in the brain): Strokes can be life threatening. Some stroke symptoms may last only minutes or a few hours and are called TIAs (Transient Ischemic Attack or mini stroke). Rapid treatment (within three hours) is vital. Medical treatments that reverse the stroke damage are available—but only if you seek immediate treatment. Stroke symptoms are:

Sudden numbness or weakness of the face, arm, or leg, especially on one side of the body

Sudden confusion, trouble speaking, or understanding speech

Sudden trouble seeing in one or both eyes

Sudden trouble walking, dizziness, loss of balance, or coordination

Sudden, very severe headache with no known cause. Can also be very severe headache that lasts for days

Clots in other locations: Muscle pain, numbness, or tingling, pale color, weakness, muscle spasm in a leg, or arm

The arm or leg feels cold, hot, or swollen to touch. May feel like a muscle strain

Extreme pain without a cause, anywhere in the body

Shortness of breath or chest pain (under the breast bone or on one side of the chest) may radiate outward from the chest. (This could indicate a clot in the lungs or a heart attack.)

Sudden heavy cough especially if you cough up blood

Rapid breathing

Heart attacks, strokes, and other blood clots need immediate medical attention. Go to an Emergency Room immediately!

If you feel these warning signs, **DIAL 911 IMMEDIATELY**. Most people wait 2 or 3 hours before seeking care, yet by then the heart may suffer significant damage. Dial 911 within 5 minutes if you or someone you are with experiences these symptoms.

~ Coping with APS ~

APS treatment is life long. Treatment of blood clots caused by APS outweighs the minimal small side effects of treatment.

Most of the time people with APS will appear on the outside just as they did before they were diagnosed. Because of this, it may be difficult for family and friends to understand that you have a life-threatening illness and that you just can't do some of the things that you could before. They can't see what is going on with your body and this can become quite frustrating on both ends.

In some cases you may want a therapist to help you adjust. Any person with a life threatening disease is at risk of being depressed. In some cases this progresses to clinical depression. It is very important to discuss your mental health with your doctor as well as your physical health.

The APS Foundation of America, Inc. has a place on the Internet where you can communicate with others who have this disease. You can find our support forum at <http://www.apsforum.com>. You are the only one who knows exactly how you feel. Others with the same disease are very understanding and support groups of some kind can make your life much fuller. Your immediate family and children should be told about your disease, how it affects you, what it means to them as well as what you may have to expect of them in support.

The good news is: many people with proper treatment live normal, full lives. Others may find their lives changed forever due to APS, but a positive attitude will mean that life can still be worthwhile and fulfilling as the correct treatment prevents further problem.

~ Other sources for APS Information ~

The ***APS Foundation of America, Inc. (APSFA)*** at <http://www.apsfa.org>. Founded in June 2005, the APS Foundation of America, Inc. is dedicated to fostering and facilitating joint efforts in the areas of education, support, research, patient services, and public awareness of Antiphospholipid Antibody Syndrome in an effective and ethical manner.

The ***APS Friends and Support Forum*** at <http://www.apsforum.com>. This is an open forum for people who have Antiphospholipid Antibody Syndrome, friends, family, and caregivers. Sometimes APS is known as APLS or APLA in the US and as Hughes Syndrome in the UK.

Please feel free to participate in any of the discussions listed, browse around, or post your own new discussion. We always welcome new members and returning members with open arms!

National Alliance Alliance for Thrombosis and Thrombophilia (NATT) at: <http://www.nattinfo.org/>. The National Alliance for Thrombosis and Thrombophilia (NATT) is a nationwide, community-based, volunteer health organization formed in August 2003. Committed to preventing and treating the array of major health problems caused by blood clots, NATT's charter members were visionary patients attending a thrombosis and thrombophilia awareness meeting at the Centers for Disease Control and Prevention. With the goal to ensure that people suffering from thrombosis and thrombophilia get early diagnosis, optimal treatment, and quality support, NATT members are committed to fostering research, education, support, and advocacy on behalf of those at risk of, or affected by, blood clots.

Rare Diseases Clinical Research Network (RDCRN) at <http://www.rarediseasesnetwork.org>. The Rare Diseases Clinical Research Network was created to facilitate collaboration among experts in many different types of rare diseases. Our goal is to contribute to the research and treatment of rare diseases by working together to identify biomarkers for disease risk, disease severity and activity, and clinical outcome, while also encouraging development of new approaches to diagnosis, prevention, and treatment.

Hospital for Specialized Surgery and Barbara Volker Center for Women and Rheumatic Disease at <http://www.hss.edu/Conditions/Antiphospholipid-Syndrome>.

Thrombosis Interest Group of Canada (T.I.G.C) at <http://www.tigc.org/default.htm>. Thrombosis Interest Group of Canada (T.I.G.C) is dedicated to furthering education and research in the prevention and treatment of thrombosis. The Thrombosis Interest Group of Canada consists of a group of 40 specialists in fields related to thrombosis who collaborate to write evidence-based or consensus-based clinical guides on the investigation, management, and diagnosis of thrombotic disorders.

~How can you help? ~

There are many ways you can help the APS Foundation of America, Inc. You can volunteer your time and talents such as in the area of fundraising, advocacy, finance, or support group experiences, donate money or purchase APS gear through our webpage at <http://www.apsfa.org> or through our Café Press at <http://www.cafepress.com/apsfoundation>.

The APS Foundation of America, Inc. is a non-profit organization. Your donations are greatly needed to help us provide awareness, support, and education of this disease. We need your enthusiasm and monetary support to help our individuals, family, friends, and caregivers battle the long-term consequences caused by APS. Thank you in advance for your support and time.

Please contact us at through our website or at 608-782-2626 for more information.

Please send donations to:

APS Foundation of America, Inc.
Post Office Box 801
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We also accept PayPal donations via our website.

We thank you for your support!

~ Disclaimer ~

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**If you think you may have a medical emergency,
call your doctor or 911 immediately.**

A team of people contributed to this publication. Information was adapted from various websites, books, and other media sources. Please contact us through the website for a complete list of sources. This document was assessed at draft stage by doctors, allied health professionals, an education specialist and people with APS. A non-medical editor rewrote the text to make it easy to understand and an APS Foundation of America, Inc. medical editor is responsible for the content overall.

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