The National Alliance
for Thrombosis
and Thrombophilia

Antiphospholipid Syndrome (APS)

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AWARENESS • PREVENTION • TREATMENT • SUPPORT

OTHER NAMES

Antiphospholipid Antibody Syndrome (APLAS)
Hughes Syndrome
Sticky Blood Syndrome
Lupus Anticoagulant Syndrome

WHAT IS IT?

People with the antiphospholipid syndrome (APS) have a tendency to form abnormal blood clots. While people need blood to clot to prevent bleeding, clotting too easily can be harmful. Blood clots may form in the arteries and block blood going from the heart to the body, or blood clots may form in the veins, and block blood going from the body back to the heart.

In pregnant women with APS, blood clots form in the placenta (the tissue that provides nutrients to the baby). This prevents nourishment and oxygen getting to the developing baby.

WHAT CAUSES IT?

Substances called antibodies defend our bodies against foreign invaders. Usually, those are viruses or bacteria. But sometimes the body gets confused. In a case of mistaken identity, it makes antibodies against itself. Diseases where the body does this are called autoimmune diseases. More well-known autoimmune diseases include lupus (SLE), multiple sclerosis, and some forms of diabetes.

An important part of our bodies are phosphorous-containing fats called phospholipids. Phospholipids and proteins that are attached to them play an important role in blood clotting. In APS, the phospholipids makes antibodies against phospholipids and these antibodies contribute to blood clot formation.

WHERE DO THE ANTIBODIES COME FROM?

In many cases, we don't know. Some people make the APS antibodies for no apparent reason. These people have primary APS. More commonly, people with autoimmune disorders, especially lupus (SLE), people with certain cancers (such as lung and ovarian), infections (like AIDS and malaria) and those taking certain medications (like the birth control pill) make APS antibodies (Asherson, Piette and Cervera, 2002). These people have secondary APS.

WHAT PROBLEMS DOES APS CAUSE?

Most importantly, it causes an abnormal blood clot. A blood clot in medical terms is called a thrombosis (two or more blood clots are called thromboses). The most common clots are deep within the legs (deep venous thrombosis or DVT). The leg becomes swollen and painful in most cases. When a blood clot breaks loose and floats away to lodge elsewhere, it is called an embolus. Blood clots from a DVT can travel to the lungs

and damage or kill parts of the lung. These are called pulmonary emboli.

If the clot occurs in an artery, it can cause a stroke or heart attack. Most strokes and heart attacks are not caused by APS, but by hardening of the arteries (atherosclerosis) in older people. Thus, younger patients with strokes and heart attacks may benefit from testing for the antiphospholipid antibodies (Hughes, 2001).

Other organs such as the kidneys, the eyes, the brain, and the liver may be affected by blood clots from APS. These clots cause different symptoms, depending on the part of the body affected. In the very worst and most rare form, catastrophic APS, multiple sites are affected at the same time. These patients are deathly ill. Half of them die (Asherson, et al., 2002).

A woman with APS may have many problems with pregnancy. She may have many miscarriages, early or late, and could have problems with high blood pressure and liver disease near her due date. The baby is also at higher risk to be born too early or too small (Balash, et al., 2002). Yet, treatments are available that may help women with APS have healthy pregnancies.

WHO GETS APS?

Typically, someone with APS is a young or middle-aged woman. APS has been diagnosed in children as young as eight months old (Carsons, 2004) or in elderly people. Men certainly can get it, but are much less likely to do so than women.

HOW DO I KNOW IF I HAVE IT?

This can be confusing. You must have had miscarriages or a clotting event and two positive laboratory tests for antiphospholipid antibodies at different times. You can have miscarriages and not have APS. You can have clots and not have APS. You can have APS antibodies and not have APS. In fact, up to 5% of healthy normal people have APS antibodies. You can have APS antibodies at one point in time only to have them disappear later.

APS antibodies can be detected by blood clotting tests for “lupus anticoagulant” or by specific antibody tests for “anticardiolipin antibodies”. The “lupus anticoagulant” or LA got its name because it was discovered in the blood of someone with lupus and it interfered with a laboratory clotting test, making it look like the person was on an anticoagulant (“blood thinner”). In fact, it is not necessary to have lupus to have the lupus anticoagulant in the body; LA causes clotting and isn’t an anticoagulant at all. Tests for LA include the DRVVT test and the LA-PTT test.

The most common types of APS antibodies are the anticardiolipin antibodies. Cardiolipin (aCL) antibodies come in three different forms (IgG, IgM, and IgA) each of which should be tested. Other less common antibodies may also be present. What role they have in APS is not clear (Misita & Moll, 2005).

The blood tests for APS antibodies are tricky to perform and are hard to compare from one lab to another. The diagnosis of antiphospholipid syndrome is difficult to make for many reasons. Doctors experienced in treating APS are best suited to make the diagnosis.

HOW IS IT TREATED?

APS is usually treated with some medication that makes it harder for the blood to clot (an anticoagulant). Although these are commonly called "blood thinners" they really don't make the blood any thinner. They work by interfering with the run-away clotting that APS causes. (See the NATT brochure "Treatment of Thrombosis" for more details.)

In general, anticoagulants fall into two categories: those that come in pill form and those that have to be injected into the skin or a vein. The only pill available now is warfarin (Coumadin®). New medications that can be taken by mouth are being developed. The injectable anticoagulants are heparin and low molecular weight heparins (LMWH).

Another way to decrease clotting is by taking medications that interfere with platelet function. Platelets are specialized blood cells that plug an injured blood vessel. Specialized clotting substances build on the plug to form the clot. Examples of anti-platelet drugs are aspirin and Plavix®. These drugs are frequently used in APS patients that have had a clot in an artery. Aspirin, by itself or with one of the injectable drugs (usually heparin) is used for pregnant women with APS. Women who have had difficulty having children because of APS have been able to have healthy babies using this treatment.

Other autoimmune diseases such as rheumatoid arthritis and lupus are treated with drugs that directly affect the immune system. These drugs can be useful in secondary and in catastrophic APS. They haven't been especially helpful in primary APS. Perhaps in the future this approach will be used.

In some severe cases, the APS antibodies are directly removed from the blood by special filters, a process called plasmapheresis.

WHAT ELSE CAN I DO ABOUT IT?

It is important to remember that many people with APS

lead fulfilling and productive lives despite the challenges of dealing with APS. There are many things that can be done to remain healthy. Some helpful tips to maintain health include:

• If you have had a DVT, wear compression socks or hose (Geerts et al., 2004). Be especially cautious when sitting for long periods of time such as on a long plane ride. In these situations, wear compression socks, drink lots of liquids (non-alcoholic), wiggle your ankles and walk frequently.

• Reduce your risk of clots from other problems, such as atherosclerosis. Work with your doctor to start a heart-healthy exercise program. Make healthy choices in foods and avoid fatty, high cholesterol meals. Don't smoke.

• Be informed! If you take Coumadin®, learn how your diet can affect your blood clotting tests (INR or protime). Be careful not to take medications or supplements that can interfere with Coumadin's effects. Your doctor or pharmacist can help you with this information.

• Avoid supplements like echinacea that stimulate the immune system.

WHERE CAN I GO FOR MORE INFORMATION?

The NATT website has current information on APS (www.nattinfo.org.

You can also get patient information from other organizations, including:

The American Auto-Immune Related Diseases Association (www.aarda.org)

APS Foundation of America, Inc. (www.apsfa.org)

Hughes Syndrome Foundation in Britain (www.hughes-syndrome.org)

National Organization for Rare Diseases (www.nord.org)

You can find many informative articles and patient support groups on the Internet by searching for "antiphospholipid syndrome" with your web search function.

Books on APS include *Sticky Blood* Explained by Kay Thackray and *Positive Options for Antiphospholipid Syndrome (APS): Self-Help and Treatment* by Triona Holden and Robert Roubey.

SELECTED REFERENCES AND FURTHER RESOURCES:

Books on APS include: Sticky Blood Explained by Kay Thackray and Positive Options for Antiphospholipid Syndrome (APS): Self-Help and Treatment by Triona Holden and Robert Roubey.

You may also wish to look at the references used to write this brochure, which include:

Asherson, R., Piette, J-C., & Cervera, R. (2002). "Primary", "secondary", "seronegative", "catastrophic" and other subsets of the antiphospholipid syndrome. In R.A .Asherson., R. Cervera, J-C. Piette, & Y. Shoenfeld (Ed), The Antiphospholipid Syndrome II: Autoimmune Thrombosis (pp.285-296). Amsterdam: Elsevier.

Balash, J. et al., 2002. Management of reproductive failure in the antiphospholipid syndrome. In R. A. Asherson, R. Cervera, J-C. Piette, & Y. Shoenfeld (Ed),), The Antiphospholipid Syndrom II: Autoimmune Thrombosis (pp.285-296). Amsterdam: Elsevier.

Carson, S., & Belilos, E. (2004). Antiphospholipid syndrome. Emedicine. Retrieved January 14, 2006 from http://www.emedicine.com/med/topic2923.htm.

Geerts, W. et al., (2004). The seventh ACCP conference on antithrombotic and thrombolytic therapy: Prevention of venous thromboembolism. Chest, 126, 3385-4005.

Hughes, G. (2001). Hughes syndrome: A patient's guide. London: Springer-Verlag.

Misita, C.P. & Moll, S. (2005). Cardiology Patient Page: Antiphospholipid antibodies. Circulation, 112, e39-e44.

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