

ANTIPHOSPHOLIPID SYNDROME

An Information Booklet



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CONTENTS

- 2 What is antiphospholipid syndrome (APS)?
- 2 History of APS
- 2 What are the main signs and symptoms?
- 4 Other features
- 4 What triggers the thrombosis?
- 5 Is APS common?
- 5 Pregnancy and APS
- 6 How is APS diagnosed?
- 7 What is the treatment for APS?
- 8 What should I do if I think I have APS?
- 8 What can I do to help myself?
- 9 The good news
- 9 Further reading
- 10 Useful addresses
- 11 Glossary

What is antiphospholipid syndrome (APS)?

Antiphospholipid syndrome (APS) is a disorder in which the blood has a tendency to clot too quickly ('sticky blood' syndrome). The clotting can affect any vein or artery in the body, resulting in a wide range of symptoms. These are described below. (The medical words in *italics* are explained in more detail in the Glossary at the end of the booklet.)

History of APS

Most of the early work leading to the detailed description of APS was carried out in the early 1980s by Dr Graham Hughes and his colleagues while studying a disease called *lupus* (see **arc** booklet 'Lupus'). In lupus the immune system goes into 'overdrive' and produces a huge variety of excess *antibodies*. While studying lupus it became clear that some patients had a tendency to form blood clots in arteries and veins. This clotting was associated with a particular antibody which 'attacks' *phospholipids* – hence the term *antiphospholipid antibodies*. It was discovered that the syndrome could exist in people without lupus. This is now known as APS or primary antiphospholipid syndrome.

What are the main signs and symptoms?

There are two main problems caused by APS. They are, firstly, blood clotting (*thrombosis*) and secondly, in women, a tendency to miscarriage (see the section

'Pregnancy and APS' below). It is important to recognise that in APS, as distinct from other clotting disorders, the thrombosis may occur in the arteries as well as in the veins – clotting in the arteries is more serious.

In APS blood clotting may affect the brain. Symptoms include headaches, memory loss/forgetfulness, slurred speech, mental sluggishness and fatigue, visual disturbances and seizures (*fits*). It is estimated that 1 in 5 people under 40 years who suffer from strokes may have APS. Some people with APS have symptoms that mimic multiple sclerosis – but there is otherwise **no** connection between APS and multiple sclerosis.

Deep vein thrombosis (DVT) of the leg is the commonest type of vein thrombosis. Apart from APS this sometimes occurs after surgery or long flights or in women taking the contraceptive pill. DVT in the leg causes pain and swelling in the calf. The whole leg may become swollen and tight. Although the diagnosis may seem obvious, it should be confirmed in hospital by tests such as ultrasound. The major concern in patients with a DVT is the risk of a small piece of blood clot travelling to the lungs (pulmonary embolism). Other internal organs, such as the kidney and liver, can also be affected by thrombosis.

APS may affect the heart. There are two main areas of the heart which can be affected: the heart valves and the coronary arteries which supply blood to the heart muscle. The heart valves may become thickened and fail to work properly. The coronary arteries may also become thicker, leading to angina.

Kidney disease in APS can cause narrowing of the blood vessels to the kidney and this may cause high blood pressure.

Other features

Some people develop a blotchy rash, often seen on the knees or wrists, with a ‘lacy’ pattern (known as *livedo reticularis*). It is an important sign in APS.

A reduction in one of the white blood cells (platelets) is also found in some people with APS.

Problems which antiphospholipid syndrome can cause

1. Thrombosis – veins (DVT)
 - arteries (stroke, high blood pressure)
 - brain (memory loss, migraine, forgetfulness, slurred speech, fits)
2. Recurrent miscarriages
3. Skin rash (*livedo reticularis*)
4. Low platelet count

What triggers the thrombosis?

There are a number of well-known factors which in general contribute to the likelihood of having a thrombosis. These include smoking, immobility (related for instance to the thrombosis seen after long flights) and the contraceptive pill. There is a slight tendency for the disease to run in families and there may be a family history of clots, of miscarriages, or of other *autoimmune diseases* such as lupus and thyroid problems.

Occasionally the thrombosis occurs during an infection such as a sore throat. However, in the vast majority of people the thrombosis comes ‘out of the blue’. In some people with thrombosis the past medical history (e.g.

migraine, recurrent miscarriages) helps to make the diagnosis of APS.

Is APS common?

APS can be seen in every branch of medicine, because it can affect a number of different organs in the body. It is increasingly recognised as a cause of disease. All age groups can be affected, from infants to the elderly. However, the majority of people with APS are aged between 20 and 50 years. It seems to affect the health of women more than men because of its effect on pregnancy.

Pregnancy and APS

As mentioned earlier, APS makes the blood tend to clot more quickly than normal (‘sticky blood’). Pregnancy itself leads to a slight increase in blood ‘stickiness’ and it is during pregnancy that many people are first diagnosed with the syndrome. The blood supply to the foetus in the womb is complicated and delicate. The presence of the antiphospholipid antibodies is particularly important during pregnancy, where the ‘sticky’ blood is unable to cross the smallest blood vessels in the afterbirth (placenta), so causing miscarriage. Although this can happen at any time during pregnancy, it is most typical between 3 and 6 months. It can also cause other pregnancy complications such as high blood pressure (*pre-eclampsia*), small babies and early deliveries. APS is now recognised as one of the most important causes of treatable recurrent miscarriages. (On pregnancy in general see **arc** booklet ‘Pregnancy and Arthritis’.)

Expert care and close monitoring of the pregnancy is essential. The success rate in APS pregnancy has now

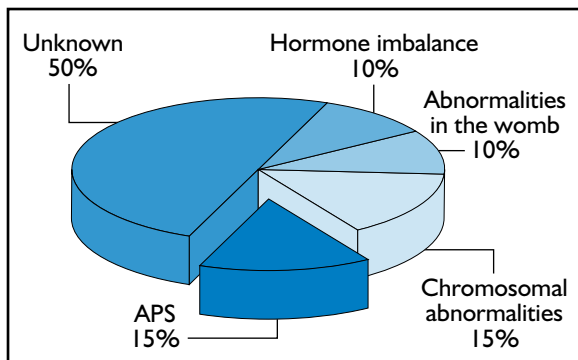


Figure 1. Causes of recurring miscarriages

risen from 20% to over 70% because of advances in the understanding and treatment of this disorder.

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

How is APS diagnosed?

There are two main blood tests that are used for diagnosis:

- the *anticardiolipin* test
- the *lupus anticoagulant* test.

Although these tests measure broadly the same thing, there are a small number of people with APS (about 20%) in whom one or other test will be negative. In other words, one test alone could miss the diagnosis, so it is important to have both. These tests are available in most major hospitals.

It is important to note that only a few individuals with antiphospholipid antibodies will actually have any problems caused by them. The level of antibodies is very important. Higher levels tend to give a much greater risk of blood clots and other APS symptoms. One positive

test for antiphospholipid antibodies does not mean that a person has APS. The test should be repeated after 6–8 weeks. A positive test on two occasions is much more important for doctors making the diagnosis than a test that is only positive once. Tests that are only just positive and that are present on only one occasion are probably not significant. This is because harmless antiphospholipid antibodies can be detected in the blood for brief periods, occasionally in association with a wide variety of conditions, including infections and certain drugs (e.g. antibiotics and certain blood pressure pills).

What is the treatment for APS?

One of the huge successes of recent medicine has been the recognition of this syndrome and the fact that treatment can prevent both the clotting complications and the miscarriages. At the moment the syndrome cannot be cured but the effects can be controlled. Treatment is with anticoagulation (blood-thinning) drugs. The three drugs most commonly used are aspirin, warfarin and heparin.

- **People with antiphospholipid antibodies but without a history of clotting** are recommended low-dose aspirin (75–100 mg) daily. Aspirin is known to make the blood less ‘sticky’. However, there are studies which suggest that some people go on to develop clots despite taking aspirin. Research is currently being done to compare the use of aspirin with very low-dose warfarin.
- **People with APS who have a history of clotting** are at risk of this happening again. This can be prevented by long-term anticoagulation treatment. Warfarin, an anticoagulant which is taken by mouth, is used and this helps to keep the blood thin. People

on treatment with warfarin have to have regular blood tests to check the intensity of the anticoagulant effect (known as INR – International Normalised Ratio). The most serious side-effect of the warfarin during treatment is bleeding. Therefore careful monitoring of the INR and warfarin dosage is very important.

- **For women with recurrent miscarriages, but without a history of clotting**, the traditional treatment is low-dose aspirin. Heparin (given by injection) is also increasingly being used – especially for people who have had previous miscarriages in mid- to late pregnancy, or other pregnancy complications such as pre-eclampsia. As warfarin is potentially toxic to the baby, a woman on warfarin who becomes pregnant will be changed over to heparin. This is less convenient for the patient, but safer for the baby. Although complications towards the end of pregnancy (for example babies born very small or early) may occur, the majority of babies do very well with no long-term problems.

What should I do if I think I have APS?

If you have one or more of the features of APS mentioned above you should seek the advice of your doctor. If necessary the appropriate blood tests can then be done, usually on two separate occasions. Your doctor may wish to refer you to a specialist – the particular kind of specialist will vary from hospital to hospital and may be a *rheumatologist* or a *haematologist*.

What can I do to help myself?

In theory, increasing the amount of essential fatty acids (EFAs) in your diet, particularly omega-3 EFAs found

in oily fish, should help reduce the risk of thrombosis, but there are no clinical trials to suggest this is the case. (See **arc** booklet ‘Diet and Arthritis’.) It is also worth noting that fish oils also contain large amounts of vitamin A, which may be harmful in pregnancy. No other ‘alternative’ treatments have been shown to help.

You should be aware of the signs and symptoms of APS and report any of these to your doctor as soon as they occur.

The good news

It is now more than 18 years since the description of the syndrome, and we now know much more about APS. More importantly, with simple blood tests diagnosis is straightforward, opening the way to early treatment. In those people in whom APS is diagnosed and treated, the outlook is very positive.

Further reading

Available from the Hughes Syndrome Foundation and major bookshops:

Hughes Syndrome : A Patient's Guide, by Dr Graham Hughes, Springer-Verlag London 2001, ISBN 9781852334574

Hughes Syndrome : Antiphospholipid Syndrome (textbook), by Dr Munther A Khamashta (Editor), Springer-Verlag London 2000, ISBN 9781852332327

The Arthritis Research Campaign (arc)

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Phone: 0870 850 5000
www.arc.org.uk

As well as funding research, we produce a range of free information booklets and leaflets. Please see the list of titles at the back of this booklet.

Hughes Syndrome Foundation

Louise Coote Lupus Unit
Gassiot House
St Thomas' Hospital
London SE1 7EH
Phone: 020 7188 8217
www.hughes-syndrome.org

An organisation specifically aimed at producing information and help for patients with APS.

St Thomas' Lupus Trust

Louise Coote Lupus Unit
Gassiot House
St Thomas' Hospital
London SE1 7EH
Phone: 020 7188 3562
www.lupus.org.uk

The charity that funds lupus research at St Thomas' Hospital and provides patient information.

LUPUS UK

St James House
Eastern Road, Romford
Essex RM1 3NH
Phone: 01708 731251
www.lupusuk.com

A support group for people with lupus and their families.

Antibody – A naturally occurring molecule produced by the body to combat infections.

Antiphospholipid antibody – An antibody which 'attacks' phospholipids (see below). Because the antibody attacks the body's own cells, rather than bacteria, it is called an autoantibody.

Anticardiolipin test – A blood test used to diagnose APS. This test measures the amount of antiphospholipid antibodies in the blood.

Autoimmune disease – A disorder of the body's defence mechanism (immune system), in which antibodies and other components of the immune system attack the body's own tissue – these are called auto-antibodies.

Chromosomal abnormalities – After fertilisation of the egg by the sperm the baby grows by multiplication of the cells. Sometimes this process of cell multiplication can cause abnormalities in the genes, called chromosomal abnormalities. These chromosomal abnormalities may be severe enough to cause a miscarriage.

Haematologist – A hospital specialist who has an interest in disease of the blood.

Livedo reticularis – A rash which occurs in APS. It looks blotchy and is seen most often on the knees and wrists. It gets its name because of its lacy pattern on the skin.

Lupus – A short term for systemic lupus erythematosus, a condition often linked to APS. (See **arc** booklet 'Lupus'.)

Lupus anticoagulant test – A blood test used to diagnose APS. This test measures the effect of the anti-phospholipid antibodies on the blood clotting time.

Phospholipids – A type of fat found throughout the body, particularly in the outer coating of cells such as the white blood cells called platelets.

Pre-eclampsia – A common condition in the second half of pregnancy in which three things occur: high blood pressure, protein in the urine, and fluid retention. Pre-eclampsia occurs more commonly in first pregnancies as well as APS.

Rheumatologist – A hospital specialist with an interest in diseases of joints, bones and muscles. As lupus is one of the conditions treated by rheumatologists, they often have an interest in APS.

Thrombosis – A blood clot which may occur in an artery or a vein.

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 Arthritis: a Guide for Teenagers
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 When a Young Person Has Arthritis

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Arthritis Research Campaign



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arc receives no government or NHS grants and relies entirely on its own fundraising efforts and the generosity of the public to support its research and education programmes.

Arthritis Today is the quarterly magazine of **arc**. This will keep you informed of the latest treatments and self-help techniques, with articles on research, human interest stories and fundraising news. If you would like to find out how you can receive this magazine regularly, please write to: Arthritis Research Campaign, Ref AT, PO Box 177, Chesterfield S41 7TQ.

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Photo courtesy of the Esbourne Gazette

Where our money goes

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This includes both basic science – trying to understand more about the disease process in order to slow down and ultimately prevent all forms of arthritis and related musculoskeletal conditions – and more clinical research aimed at helping to improve the daily lives of patients.





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