

Hughes Syndrome  
(antiphospholipid syndrome)  
- clinical features

General features

Hughes (antiphospholipid) Syndrome or APS is characterised by an increased tendency to blood clotting, both in arteries and veins. The clotting can affect many parts of the body and can cause recurrent miscarriage – please refer to the Pregnancy and Fertility leaflet for more information about this important aspect of the condition. Many patients with antiphospholipid antibodies (aPL) in their blood stream will not suffer any adverse effects, but the most common symptoms are headaches, fatigue, cold circulation (including 'blotchy' skin) and memory problems. As Hughes Syndrome is an autoimmune condition, it is not uncommon to find other autoimmune disorders such as thyroid disease, arthritis, pernicious anaemia or lupus in other members of the family.

Arm and leg clots - DVTs

The commonest limb vein clot is in the leg – acute, painful swelling often in the calf, but they can also develop in the arm. This can happen spontaneously, or can be precipitated by other factors, such as long journeys, operations and the contraceptive pill. The main danger of a limb deep vein thrombosis (DVT) is the spread of the clot to the lung.

Lung clots

Clots in the legs, arms or pelvis can become dislodged and end up in the lung where they are known as pulmonary emboli. A lung clot can be dramatic and life threatening, with sudden, acute chest pain, leading to severe shortness of breath, collapse or even death.

Skin and Circulation

The cold circulation is often seen on the skin as 'livedo reticularis' – a blotchy, lacy pattern of blood vessels, most commonly seen on the knees, thighs and upper arms. In some it can be quite widespread. The poor circulation (especially where there has been a vein thrombosis) can occasionally lead to ulceration, often

in the lower leg. Bruising is also very common (particularly for patients taking aspirin) and is sometimes a clue to low platelet count in the blood.

Brain

The clinical manifestations of impaired circulation to the brain are very diverse and range from headache, balance problems and mild memory loss through to seizures and even stroke. More details about this very significant aspect of Hughes Syndrome are provided in a separate leaflet.

Heart

The classical feature of an impaired blood supply to the heart is angina. Most commonly brought on by exercise, stress or the cold, it can be quite subtle,

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generally improving in Hughes Syndrome when anticoagulants are started. An entirely different heart problem sometimes seen in Hughes Syndrome is valve disease. Blood clots occasionally gather around the delicate heart valves and can lead to malfunction. As effective anticoagulation treatment halts this process, early diagnosis using echocardiography is important.

#### Kidney

Clotting or sludging of blood in the kidney and its blood vessels can lead to raised blood pressure and impaired kidney function.

#### Liver and digestive system

That a degree of blood clotting can affect the liver is suggested by the common finding of mildly abnormal liver function tests on blood analysis. More serious untreated blood clotting can lead to a rare liver condition known as Budd-Chiari Syndrome. Early diagnosis and treatment generally leads to improvement of the milder forms of liver involvement.

#### Eye

Thrombosis in the blood vessels of the eye can lead to a devastating loss of vision in one eye. Other manifestations can include a partial loss of vision, and the flashing lights and zig-zag patterns of the migraine attack.

#### Blood

Some patients with Hughes syndrome develop a low platelet count (e.g. 50 – 90,000: the 'normal' range being over 140,000). This can cause easy bruising and even bleeding. It is hard to envision bleeding as a (rare) feature of what is essentially a clotting disorder. Fortunately most cases respond to a course of steroids - see the Treatment Leaflet for further information.

#### Bones and joints

Obstruction of blood flow to the head of the femur can lead to a collapse of the hip joint known as 'avascular necrosis'. Less commonly this can happen in other joints. Recently, we have also reported that bone fractures (e.g. in the foot), can be seen in the syndrome – possibly again as a result of impaired blood supply to the area.

#### Pregnancy

In pregnancy, the blood is slightly 'stickier'. It is therefore not surprising that Hughes Syndrome is often first manifest or diagnosed in pregnancy, sadly usually after a series of miscarriages. The topic of pregnancy and APS is discussed in a separate leaflet.

#### Catastrophic APS

Very occasionally, a dramatic series of widespread blood clots causes life threatening involvement of the brain, adrenals, lungs and heart. The reasons for the sudden acceleration of the disease (almost invariably

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in untreated or under-treated patients) are, at the  
present time, unclear, though infection is suspected in  
some cases.

If you would like to learn more about  
Hughes Syndrome, we have a number of  
excellent books and guides available:

Hughes Syndrome: A Patients Guide -  
£6.00  
Living with Hughes Syndrome - £7.00  
Sticky Blood Explained - £8.00  
More Sticky Blood - £7.99  
The Big 3 (GP Booklet) - £2.00

These books and other merchandise can  
be purchased from our website:  
[www.hughes-syndrome.org](http://www.hughes-syndrome.org)

Charity Office

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