

# LUPUS and Blood Disorders



## THE LUPUS UK RANGE OF FACT SHEETS

Further fact sheets are available as follows:

- LUPUS Incidence within the Community
- LUPUS A Guide for Patients
- LUPUS The Symptoms and Diagnosis
- LUPUS The Heart and Lungs
- LUPUS and the Brain
- LUPUS and the Kidneys
- LUPUS The Joints and Muscles
- LUPUS The Skin and Hair
- LUPUS The Mouth, Nose and Eyes
- LUPUS and the Feet
- LUPUS Fatigue and your Lifestyle
- LUPUS and Men
- LUPUS and Light Sensitivity
- LUPUS and Pregnancy
- LUPUS and Medication
- LUPUS and Associated Conditions

LUPUS UK is the registered national charity caring for people with presently incurable lupus and has over 5,500 patients in membership who are supported by the Regional Groups.

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Please contact our National Office should you require further information about lupus. LUPUS UK will be pleased to provide a booklist and details of membership.

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## What else does the antiphospholipid syndrome cause?

Apart from causing low platelet counts, deep vein thrombosis and possibly pulmonary embolism, antiphospholipid antibodies can cause blood clots in the major arteries of the body. This can result in conditions such as heart attack or stroke. In women of childbearing age, these antibodies can also cause miscarriages or stillbirths due to blood clots forming in the placenta which prevent nutrients from reaching the developing baby during pregnancy.

## How is antiphospholipid syndrome diagnosed?

Antiphospholipid syndrome is diagnosed by finding antiphospholipid antibodies in the blood and clinical features of increased blood clotting (for example recurrent deep vein thromboses or miscarriages). The antibodies can be detected by a specific test for the type of antibody, such as anticardiolipin antibodies, or by abnormalities in conventional clotting tests. Antiphospholipid antibodies interfere with many of the conventional clotting tests because these tests depend on phospholipid for the test to work normally. If antiphospholipid antibodies are present, they prevent the blood from clotting, making it look as if there is a shortage of clotting factors. However, in reality the antibody acts as an inhibitor of the clotting test and in real life this is associated with an increased risk of clotting (this paradox is confusing to everyone). If you mix the serum part of the blood containing the antibodies with serum from a healthy person, the antiphospholipid antibodies still interfere with the clotting test, whereas if there is a clotting factor deficiency (as in haemophilia), the mixing results in correction of the missing factor and the clotting test becomes normal. This is the basis for some special tests of blood clotting used to diagnose antiphospholipid syndrome.

# LUPUS and Blood Disorders

Systemic lupus erythematosus (SLE or Lupus) has many clinical and laboratory manifestations. This article relates to those manifestations which concern the cells and clotting factors that circulate in the blood. As with other manifestations of lupus, the underlying cause is usually the presence of antibodies which are proteins that recognize and bind to other proteins in the body. However, instead of getting rid of an unwanted foreign protein (which is what normal antibodies do), the antibodies in lupus recognize self-molecules (usually proteins) and interfere with the normal function of our own proteins and the cells containing the proteins.

## What is blood?

Blood is made up of cells and a protein rich liquid containing many substances including antibodies (serum). The cellular part is composed of red cells (which contain the oxygen carrying molecule haemoglobin), white cells (which fight infection and can be subdivided into several types, of which the neutrophils and lymphocytes are particularly important) and platelets (which are involved in clotting, the process which stops us bleeding when we cut ourselves). Clotting also involves a number of special proteins called clotting factors in addition to the platelets. A full blood count test measures the number of red and white cells, platelets and the amount of haemoglobin circulating in the blood. There are special tests for assessing the clotting properties of blood (see below under “how is antiphospholipid syndrome diagnosed”)

## Why do lupus patients have low white cell counts and what is the significance of this?

In SLE, antibodies directed against white cells are very common. A lower than normal lymphocyte count is found on the full blood count in about 95% of lupus patients. This is due to the presence of antibodies to lymphocytes which results in the destruction of the antibody-coated lymphocytes. Fortunately, this rarely causes a clinical problem because more lymphocytes are released from the bone marrow where they are made. This means that there are enough lymphocytes to fight infection, especially those due to viruses like influenza (“flu”). However, high doses of certain drugs can also cause destruction of lymphocytes and, because the bone marrow where these cells are made is also attacked by such drugs, there may not be enough lymphocytes to fight infection. So in this case there is an increased risk of infections, especially viral infections. Cyclophosphamide, which is sometimes used to treat more severe forms of lupus such as kidney disease, can have this effect. Consequently the white cell count is regularly checked in people receiving this drug so that the drug dose can be adjusted if necessary.

Low neutrophil counts are less common than low lymphocyte counts as a result of lupus and is more common in people of Afro-Caribbean origin (even in the absence of lupus). Low neutrophil counts can also be the result of drugs such as cyclophosphamide and azathioprine and if severe can be associated with bacterial infections such as pneumococcal pneumonia. Again, regular blood monitoring tests will usually prevent this complication by allowing the drug dose to be adjusted before a problem occurs.

## Why do lupus patients become anaemic and what effect does this have?

Anaemia means that there are less red cells (and therefore less haemoglobin to carry oxygen) in the

blood than there should be. Low red cell counts and the associated low level of haemoglobin in the blood can result from the effect of antibodies attacking the red cells and causing their destruction, a process called haemolytic anaemia. It is more common though to have anaemia due to poor production of red cells in the bone marrow. This usually occurs as a side-effect of general inflammation in the body due to lupus. Rarely is anaemia caused by drugs but this is possible. Whatever the underlying cause of anaemia, the end result is to cause the person to feel tired (fatigue) in proportion to the loss of red cells (haemoglobin), although there are other (often less well understood) causes of fatigue in lupus. In more severe cases, the person may become short of breath even in the absence of lung disease because there is not enough oxygen in the blood.

## What are the effects and causes of low platelets in lupus patients?

Low platelets are usually due to antibodies and less commonly the result of drug side-effects. When the count is very low there is an increased risk of bruising and bleeding; fortunately this is a rare manifestation of lupus. Surprisingly there is another type of antibody that can reduce the platelet count (usually only mildly), so there is no bleeding but instead these antibodies can interfere with platelet function and this causes increased clotting, known as thrombosis. This is a painful condition, as not enough blood can get through a blood vessel containing a clot and this causes damage to that part of the body. This can occur, for example, in the calf (deep vein thrombosis), and the clot can spread from a leg vein through the circulation to the lungs (pulmonary embolism). The underlying cause of this thrombotic condition is known as antiphospholipid (or Hughes) syndrome, as there is a group of antibodies present which interferes with phospholipids in platelets and other cells and proteins involved in clotting.