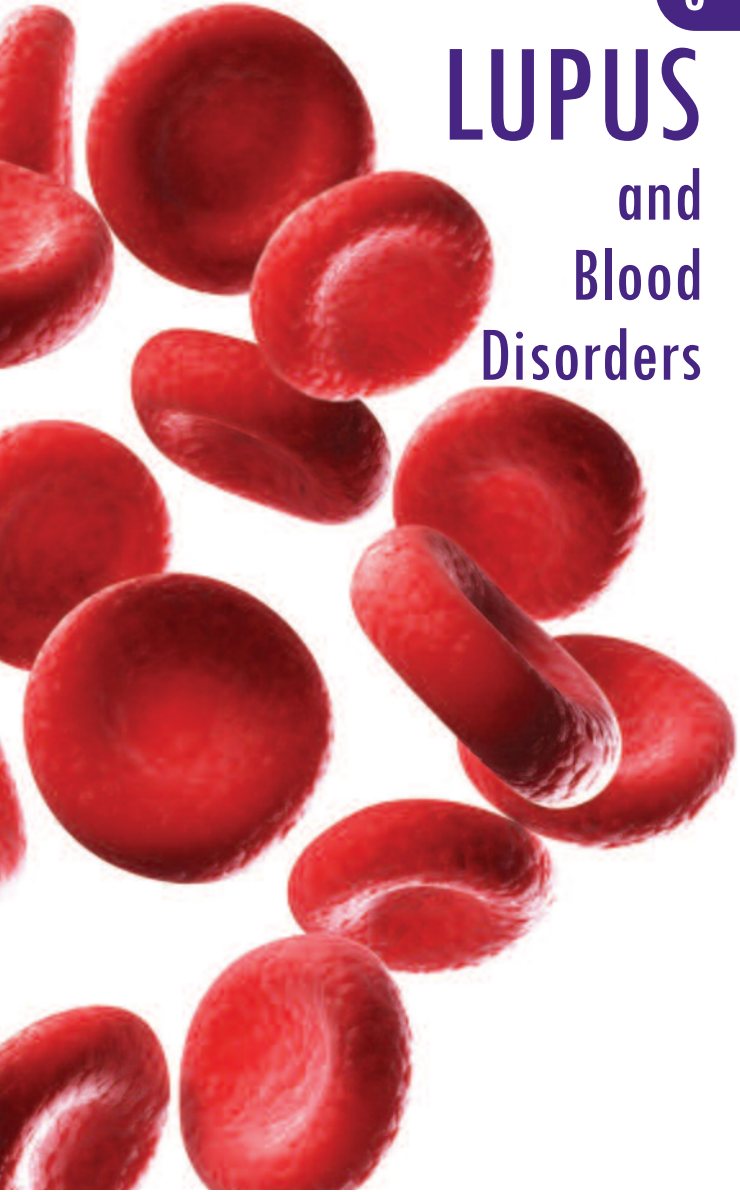


LUPUS

and
Blood
Disorders



LUPUS and Blood Disorders

Systemic lupus erythematosus (SLE or lupus) has many clinical and laboratory manifestations. This factsheet relates to those described as ‘haematological manifestations’ of the disease, which primarily involves the effects of the disease on the cells and clotting factors that circulate in the blood.

What is blood?

Blood is made up of cells and a protein rich liquid called serum, which contains many substances, including antibodies. Antibodies protect against infection but in lupus they behave differently and become confused resulting in them attacking otherwise healthy tissue. 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. In this case there is an increased risk of infections, especially viral infections. Immune suppressing drugs which are often 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 these drugs 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 are 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 azathioprine. A low neutrophil count may also be seen between 1-2 weeks after treatment with cyclophosphamide (a drug used to treat more severe forms of lupus). If there is a severely low neutrophil count this can increase the risk of 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. This is commonly seen in people with lupus with the majority of patients reporting this at some time during their illness. Low red cell counts and the associated low level of haemoglobin in the blood can result from the effect of the antibodies seen in lupus attacking the red cells and causing their destruction, a process called haemolytic anaemia. A more common cause of anaemia is due to a lack of 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. There are a number of other common causes of anaemia that are also seen in people who do not have lupus such as anaemia due to bleeding or low levels of important vitamins such as iron, vitamin B12 and folate. These are often not the direct result of lupus but are another vital cause to consider. 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. Sometimes, they may also describe palpitations (the sensation of the heart beating quickly or skipping/missing a beat).

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 blood to clot, known as thrombosis. These clots can occur in different blood vessels throughout the body and will often cause pain, 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.





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 together with the typical clinical features of increased blood clotting (for example recurrent deep vein thromboses or miscarriages). The antibodies can be detected by a specific test such as anticardiolipin antibodies, or by abnormalities in conventional clotting tests, rather confusingly known as lupus anticoagulant. These antibodies need to remain positive and therefore a truly positive result requires the test to be repeated at an interval of 12 weeks to confirm their ongoing presence.



Why do some lupus patients have low blood proteins?

The levels of blood proteins such as albumin can be low due to chronic inflammation. A low albumin is also commonly seen when the kidney leaks protein in the urine when it is inflamed in lupus (nephritis). This can cause leaking of fluid from the blood vessels into the surrounding tissues and can cause swelling (in particular in the ankles and legs). Low total levels of the normal protective antibodies (immunoglobulins) may be due to long term use of immune suppressing drugs including antibody treatments like rituximab. This may increase the risk of infections. The levels of these immunoglobulins will be checked regularly, in particular in anyone who has undergone or will undergo treatment with rituximab.

The LUPUS Range of Factsheets

A range of factsheets is available as follows:

1. LUPUS Incidence within the Community
2. LUPUS A Guide for Patients
3. LUPUS The Symptoms and Diagnosis
4. LUPUS The Joints and Muscles
5. LUPUS The Skin and Hair
6. LUPUS Fatigue and your Lifestyle
7. LUPUS and Pregnancy
8. LUPUS and Blood Disorders
9. LUPUS and Medication
10. LUPUS and the Kidneys
11. LUPUS and Associated Conditions
12. LUPUS and the Brain
13. LUPUS The Heart and Lungs
14. LUPUS The Mouth, Nose and Eyes
15. LUPUS and Light Sensitivity
16. LUPUS and the Feet
17. LUPUS and Men
18. LUPUS and Mixed Connective Tissue Disease
19. LUPUS Bone Health and Osteoporosis

LUPUS UK is the registered national charity caring for people with presently incurable lupus and has over 5,000 members who are supported by the Regional Groups.

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Please contact our National Office should you require further information about the sources used in the production of this factsheet or for further information about lupus. LUPUS UK will be pleased to provide a booklist and details of membership.



St James House, Eastern Road, Romford, Essex RM1 3NH

Tel: 01708 731251 www.lupusuk.org.uk

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