

## **Lupus – A Guide for Patients**

Lupus is a disease where the immune system becomes overactive. Antibodies are produced and almost all organs of the body can be involved.

### **Who gets lupus?**

Lupus is most common in young females (teens, 20s and 30s). Men and children can also have lupus. The ratio of females to males is nine to one.

### **Is it a world-wide disease?**

Yes, lupus affects people from all countries in the world. There are certain countries in which the disease appears to be more prevalent, for instance the Caribbean, the Far East and China.

### **What is the outlook?**

Most patients with lupus can expect a normal life-span. The disease, if diagnosed early and treated appropriately at an early stage, most commonly settles and ultimately goes into remission - i.e. the patient requires no medication.

### **What are the features of lupus?**

Fatigue, flu-like illness, skin rashes (including the classical 'butterfly' rash on the cheeks and nose), hair-loss and, more importantly, internal organ involvement including pleurisy, kidney disease and brain inflammation. Some patients with lupus have a clotting tendency and this can present, for instance, as a thrombosis in the vein or an artery.

### **How is it treated?**

Early in the disease, usually in the more aggressive stages, it is treated with corticosteroids and/or antimalarials. Where there is kidney or brain disease other drugs such as immunosuppressives are used. In those patients with a clotting tendency anticoagulants are used. Management is usually aimed at reducing medication to the lowest dose possible and, ultimately, to weaning the patient off stronger medication.

### **How can I help myself?**

Some of the factors which exacerbate lupus are stress, excessive sunlight and, occasionally, drug allergies (especially to the antibiotic Septrin). Once the patient is treated there is every possibility of a normal lifestyle. Even those patients who are sensitive to ultraviolet light and develop skin rashes can, with normal commonsense protection against excess UV exposure, lead a reasonably normal lifestyle. There are no diets which have been found especially helpful in lupus though it must be said that some patients do appear to be allergic to certain foods and the only way to find this out is by trial and error.

### **Where can I find out more?**

In addition to the self-help societies such as **LUPUS UK** there are patient networks in most countries of the world.

## **Symptoms and Diagnosis**

### **Fatigue**

This is one of the most common and certainly one of the most prominent features of lupus. Patients often describe it as an unnatural fatigue. Its causes are not well understood. Often it precedes the diagnosis by months or years and only when treatment has been successfully started does the patient realise how major a feature it had been.

### **Aches and pains**

The majority of lupus patients suffer at some stage from joint and muscle pains. In many patients this presents as 'pain all over'. In acute flares of lupus the symptoms are often described as being 'flu-like'. Unlike other rheumatic diseases such as rheumatoid arthritis, there is often very little to see in the way of joint swelling. It is not just the joints that are affected but the tendons and muscles as well. In the majority of cases the joint inflammation does not progress to permanent damage.

### **Fevers**

Fever is usually a feature of a flare of the disease. Fever is unusual when the disease is in a quiet phase: thus in an adult or a child known to have lupus who develops fever the possibility that a separate diagnosis - infection - might be present always needs consideration.

### **Rashes**

A wide variety of skin rashes occur in lupus. Traditionally these are sun-sensitive (photosensitive) but this is not always the case. The commonest rashes are on the cheeks (the butterfly rash across the nose and cheeks), on the elbows, on the palms and soles and on the V-neck area. The rashes vary from pinkish discolouration through to blisters and small pinpoint 'blood spots' (purpura). Most rashes in lupus have a tendency to come and go.

### **Hair loss**

Hair loss is one of the most important features of active lupus. It may be the first manifestation of the disease and is often first noticed by the patient as hair on the pillow. In some cases hair loss is patchy and even extreme. Fortunately, in the vast majority of patients the hair re-grows after successful treatment, though hair regeneration is often notoriously slow.

### **Headaches**

Headaches are a major feature of lupus. In some patients a history of headaches or a typical migraine go back to the patient's teens and pre-date the diagnosis by many, many years. There is almost certainly a variety of causes of headaches in systemic lupus. One specific and important cause is 'sticky blood' caused by the presence of antiphospholipid antibodies.

## **Depression**

Depression is an important feature of lupus. It is sometimes simply attributed to 'being unwell' or having tiredness and pain. However, in many patients it is far more important than this and is a primary feature of the disease. It sometimes responds well to management of the lupus itself and is clearly a central feature of the lupus process. In some patients the return of depression is a tell-tale sign that the lupus is flaring.

## **General symptoms**

As almost every organ in the body may be affected at some time, the symptoms and signs are legion and can include irritation of the eyes (sometimes associated with dry eyes), mouth ulcers, chest pain (pleurisy is, for example, important in active lupus), weight loss and ankle swelling.

## **Diagnosis**

The diagnosis of lupus is usually made on clinical grounds. The combination of some of the features described above, especially the skin rashes, usually but not always makes the diagnosis clear. Unfortunately, in many patients, especially those who do not have the classical tell-tale rashes, the diagnosis is missed. This is particularly true for those with more 'vague' symptoms such as fatigue, depression or headaches. Often the patients are given the wrong diagnosis such as ME or 'atypical multiple sclerosis'. Diagnosis is critical and any individual in whom lupus is suspected (or for that matter the relative or offspring of any individual with lupus in whom the diagnosis is a consideration) should have the simple blood tests performed.

## **Lupus blood tests**

Lupus is now almost invariably diagnosed by blood tests. These invoke a small amount of blood and are extremely sensitive. There are five major blood tests carried out on the blood sample.

### **a. Antinuclear antibody (ANA)**

This cheap and reproducible test is the 'screening test' for lupus, being positive in over 90% of cases. It is not specific for lupus but because of its simplicity is a useful first step in diagnosis.

### **b. DNA antibodies**

This is the highly specific test for lupus. For some unknown reason the presence of antibodies against double-stranded DNA is the hallmark of lupus. It is very specific for this disease and rarely found in any other condition. Strongly positive anti-DNA antibody tests provide almost total proof of the diagnosis. The level or titre of the antibodies provides a rough guide to disease activity and is used by physicians to monitor the ups-and-downs of the disease.

### **c. ENA**

The term 'extractable nuclear antigens' applies to a battery of other antibodies which are found in lupus variants such as Sjogren's syndrome and mixed connective tissue disease.

#### d. Antiphospholipid antibodies

These tests are associated with the important problem of 'sticky blood'. Patients with high levels of antiphospholipid antibodies have an increased tendency to clotting both in the veins and arteries, and in pregnant women with these antibodies there is a risk of thrombosis of the placenta leading to miscarriage. It is now recognised that many women with recurrent miscarriages have antiphospholipid antibodies and that successful pregnancies are possible when the patient with sticky blood is treated either with aspirin or with an anticoagulant.

#### e. Complement

This is a term used for a group of proteins in the blood which are involved in the immune process. In active lupus the levels of complement (usually measured as C3 and C4) are low and these often provide a clue to the degree of disease activity.

#### **General blood tests**

In addition to the specific blood tests, the physician usually requests a full blood count and biochemistry. The blood count in lupus can show low white cells, low red cells and low platelet counts. Biochemical tests are important, especially the creatinine and urea which are raised if there has been evidence of kidney disease. Two blood tests, the ESR and the C-reactive protein (CRP) are used as barometers of disease activity.

#### **Urine tests**

Testing the urine is vital in lupus patients and it is the practice in some lupus clinics to teach all patients how to test their own urine. The simple test uses a 'dip-stick' to check for protein - often the earliest clue to the presence of kidney disease. More precise urine tests are performed on a MSU (mid-stream urine - a sample of urine sent to the laboratory for microscopic analysis). Under the microscope, the presence of white cells, red cells or clumps of cells (casts) is recorded - all possible signs of kidney disease. Finally, all urine sent to the laboratory is tested for bacterial infection.

#### **More complicated tests**

The lupus patient may require specialised tests to look for more widespread organ involvement. These will include echocardiograms, brain scan (NM), kidney scans and, if there is evidence that the kidney is inflamed, possibly a kidney biopsy. Having said this, for the majority of lupus patients attending routine lupus clinics, a simple blood test and urine test are the basic requirements. From these two analyses a broad picture of the degree of lupus activity can be readily obtained.