

The Diagnosis of Lupus



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LUPUS UK is the national charity caring for those with systemic lupus erythematosus (SLE) and discoid lupus erythematosus (DLE) supporting our members through the Regional Groups and advising others as they develop the symptoms prior to diagnosis

You can help by taking up membership.
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THE DIAGNOSIS OF LUPUS

Lupus is an autoimmune disease, a type of self allergy, whereby the patient's immune system creates antibodies which instead of protecting the body from bacteria, viruses and other foreign matter attack the person's own body tissue.

This causes symptoms of extreme fatigue, joint pain, muscle aches, anaemia, general malaise, and can result in the destruction of vital organs.

It is a disease with many manifestations, and each person's profile or list of symptoms may be different. Lupus can mimic other diseases, such as multiple sclerosis and rheumatoid arthritis and diagnosis is often delayed.

Physicians are often cautious with the diagnosis as they do not want to label anyone until they are certain of lupus. Moreover, a careful review of the patient's entire medical history is necessary, coupled with analysis of results obtained from tests relating to the immune status to provide accurate diagnosis.

Lupus is a complex disease in which almost every system in the body can be affected, and the diagnosis is based on a combination of symptoms, signs and test results. Once a diagnosis of lupus is made, the patient's symptoms should be treated as necessary. The goal of the treatment is to control the symptoms and the disease so that the patient can lead as normal a life as possible.

Professor Graham RV Hughes MD FRCP
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DIAGNOSIS OF LUPUS or any other chronic illness may be established using the 5 Step Programme.

1. **Review patient symptoms**
2. **Detailed physical examination**
3. **Battery of tests**
4. **Rule out other diseases**
5. **Time is sometimes necessary to observe the course of the disease.**

THE FIRST PRINCIPLE in making a diagnosis of lupus is that the individual has clinical evidence of a multi-system disease, and several manifestations such as those listed below may be present:-

SKIN	Rashes, mouth ulcers, hair loss
JOINTS	Pain, sometimes swelling
KIDNEY	Abnormal Urinalysis (especially protein in the urine)
LINING MEMBRANE	Pleurisy, Pericarditis, Peritonitis
BLOOD	Low white cells, Anaemia
LUNGS	Shortness of breath, cough
NERVOUS SYSTEM	Headache, phobias, memory problems

THE SECOND PRINCIPLE is to examine the status of the immune system. The most useful test is the anti-DNA antibody test. Testing for anti-phospholipid antibodies (aPL) is important.

The onset of lupus can be gradual with new and different symptoms appearing over weeks, months or even years. The symptoms are often hard to describe and can come and go suddenly, therefore it may often be that the patient might begin to feel “it is all in the mind”. As a consequence such patients are sometimes categorised as hypochondriacs.



The symptoms of lupus seem to fall into two categories, non-specific and specific.

NON-SPECIFIC SYMPTOMS

1. **Fatigue -** the most frequent symptom that affects people with lupus
2. **Weight loss**
3. **Aches & pains**
4. **Fever -** indication that lupus is becoming active
5. **Swollen Glands**

Other additional problems commonly experienced by patients may be headaches, rashes, increase in hair loss, miscarriage and poor circulation.

SPECIFIC SYMPTOMS

To help distinguish lupus from other diseases, physicians of the American Rheumatism Association have established a list of 11 abnormalities which, when combined, point to lupus.

To make a classification of lupus the patient must have had at least FOUR of these 11 manifestations at any time since the onset of the disease.

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|---------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------|
| 1. MALAR RASH | Fixed red rash over the cheeks |
| 2. DISCOID RASH | Red patches of skin associated with scaling and plugging of the hair follicles |
| 3. PHOTSENSITIVITY | Rash after exposure to sunlight |
| 4. ORAL ULCERS | Small sores that occur in mucosal lining of mouth and nose |
| 5. SEROSITIS | Inflammation of the delicate tissues covering internal organs, and abdominal pain |
| 6. ARTHRITIS | Very common in lupus, usually pain in the joints and tendons |
| 7. RENAL DISORDERS | Usually detected by routine blood and urine analysis |
| 8. NEUROLOGICAL DISORDER | Seizures or psychosis, balance problems |
| 9. HAEMATOLOGICAL DISORDER | Haemolytic Anaemia, Leukopenia, Thrombocytopenia |
| 10. IMMUNOLOGIC DISORDER | Tests anti-DNA, anti-SM antibodies, antiphospholipid antibodies |
| 11. ANTI-NUCLEAR ANTIBODY (ANA TEST) | When found in the blood and the patient is not taking drugs, it is known to cause a positive test for lupus in most cases, but is not necessarily conclusive |

DNA ANTIBODIES are highly specific for lupus. Presence of antibodies against double-stranded DNA is the hallmark of lupus 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 antibodies provides a rough guide to disease activity.

SIGNS AND SYMPTOMS

which may signal that a lupus flare is beginning

Be aware of one or more of these symptoms:

-  Persistent fatigue out of proportion with what you would usually expect
-  Persistent weakness
-  Aching all over
-  Fever, which may be slight to high (you can check your temperature yourself)
-  Persistent loss of appetite
-  Involuntary weight loss
-  Recurring nose bleeds
-  Increasing hair loss
-  Unexplained skin rash anywhere on the body
-  Hives
-  Sores on the skin
-  Painful joint(s)
-  Stiffness of the joints when waking up in the morning
-  Chest pain which increases with breathing
-  Shortness of breath
-  Persistent unusual headache
-  Recurring or persistent abdominal pain
-  Persistent, increasing swelling of the feet and legs
-  Puffy eyelids
-  Blood or protein in the urine

Lupus can be a repetitive disease: watch for recurrence of symptoms from when your disease started.

AND HELPFUL HINTS...

The American Rheumatism Association criteria has provided the benchmark for the classification of lupus for the last 20 years. Many thousands of lupus patients passing through St Thomas' Hospital led Prof. Graham Hughes to offer the following 14 criteria aimed more towards diagnostic help and not to classification.

1. Teenage 'growing pains'

Growing pains, at least in the UK, is a label widely used for joint pains in teenagers and seems to cover a spectrum of rheumatology from arthritis variants through to lupus.



2. Teenage migraine

Headache, cluster headache and migraine can be encountered and a strong history of teenage migraine may be of lupus significance, either at that time or subsequently.

3. Teenage 'glandular fever'

Prolonged teenage glandular fever is a label which crops up time and time again in lupus patients and prolonged periods off school in many SLE patients is a recurrent theme.

4. Severe reaction to insect bites

This is a feature of so many lupus patients. Not only are they susceptible to insect bites but often reactions are severe and prolonged – the skin is a major organ affected by lupus.



5. Recurrent miscarriages

Lupus itself seems not to be a cause of recurrent miscarriage but where the antiphospholipid syndrome (APS) is present, recurrent spontaneous fetal loss is/can be significant.

6. Premenstrual exacerbations

Although difficult to quantify, it is believed that significant premenstrual disease flare is sufficiently prominent in lupus to be included in this list. All rheumatic diseases are clinically influenced by the menstrual cycle.

7. Septrin (and sulphonamide) allergy

Adverse reactions to these drugs is quite common in lupus and the clinical onset of the disease may have coincided with the use of eg Septrin.

8. Agoraphobia

Agoraphobia/claustrophobia are often present at a time when lupus disease is active. A history of these conditions can be protracted, lasting for months or even years. In many cases the history is not volunteered or the episodes are in the interim considered unrelated to lupus.

9. Finger Flexor Tendonitis

Arthralgia and tenosynovitis are common features in lupus and although not specific, the finding of mild to moderate ten-finger flexor synovitis is a useful pointer in the presence of other lupus features. It is subtly yet significantly different in pattern from other arthritic diseases.

10. Family history of autoimmune disease

As the genetics and statistics of the various autoimmune diseases become better defined, the strength of a particular family history will become more precise. The family history is important, as lupus may be genetically determined.



11. Dry Shimer's Test

A 'bone dry' Shimer's test (levels of eye moisture) points towards one of the autoimmune diseases and in the patient with vague or nonspecific symptoms is worth its weight in gold.

12. Borderline C4

Genetic complement deficiencies have been known to be associated with lupus for over three decades and in the diagnostically difficult patient, especially where a family history is present, repeated borderline C4 levels can be significant indicators.

13. Normal CRP and raised ESR

An important diagnostic aid. A very low CRP in an otherwise inflammatory situation is strongly supportive of lupus or primary Sjogren's syndrome.

14. Lymphopenia

In the patient with non-specific complaints and unremarkable blood tests, a borderline or low lymph count can be overlooked. It can be common in lupus and is certainly worth inclusion among minor criteria.

LUPUS UK

LUPUS UK is a registered charity with more than 5,500 patient members supported by the Regional Groups. We provide a large variety of information for those seeking a better understanding of lupus.

The charity offers a comprehensive range of books for purchase and can supply free of charge Information Packs for individuals, nurses and GPs. The LUPUS UK national magazine "LUPUS UK News & Views" is published three times during the year with particular emphasis on medical articles concerning lupus.



Publicity materials, leaflets, posters, a dvd for the newly diagnosed, media releases and more are always available from the charity's National Office for better awareness about lupus in clinics, hospitals and public places.