

# Clinical features of patients with systemic lupus erythematosus (SLE) attending the SLE outpatient clinic at Universitas Hospital in Bloemfontein, South Africa

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## Abstract

**Background:** Systemic lupus erythematosus (SLE) is an autoimmune disease, a type of self-allergy, whereby the patient's immune system creates antibodies that attack the person's own body tissues instead of protecting the body from bacteria and viruses. In most cases the cause of SLE is unknown, although it is believed that many factors may be involved, including genetic predisposition and environmental factors such as excessive sun exposure, infections, antibiotics (especially those in the sulpha and penicillin groups), extreme stress, certain drugs, and hormones. Currently, there is no single laboratory test that can determine whether a person has lupus or not. To assist the physician in the diagnosis of lupus, the American College of Rheumatology (ACR) has compiled a list of 11 symptoms or signs of which a person should have four or more to be classified as SLE. The genetic pool and environmental factors differ in different regions. The Free State and Northern Cape are known for a dry and sunny climate, as well as cold winters. The aim of this study was to determine the most common features of patients with systemic lupus erythematosus attending the outpatient clinic at Universitas Hospital in Bloemfontein, South Africa.

**Methods:** For this descriptive study, the study population included all patients attending the SLE clinic at Universitas Hospital diagnosed with SLE according to the ACR classification criteria. Patients were only included if they had at least one follow-up visit. Patients who had discoid lupus were excluded. Data were collected from patient files using a confidential and anonymous data form.

**Results:** Data were obtained from 76 patients: 71 females (94.7%) and five males (5.3%). African patients accounted for 61.3% of the study population, whites for 33.9%, Asians for 1.6% and coloureds for 3.2%. Patients most frequently had immunological (90.8%), mucocutaneous (86.9%), musculoskeletal (85.5%) and cardiovascular problems (77.6%).

**Conclusion:** Most of the findings correlate with similar studies worldwide. However, mucocutaneous manifestations and Raynaud's phenomenon were more prevalent in our study population. From this it can be deduced that the climate may play an important role. Further research needs to be conducted to investigate this hypothesis.

Ⓟ This article has been peer reviewed. Full text available at [www.safpj.co.za](http://www.safpj.co.za)

SA Fam Pract 2008;50(1):68

## Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease, a type of self-allergy, whereby the patient's immune system creates antibodies that attack the person's own body tissues instead of protecting the body from bacteria and viruses.<sup>1</sup> This causes symptoms of extreme fatigue, joint pain, muscle aches, anaemia and 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 is different. SLE can mimic other diseases, such as multiple sclerosis and rheumatoid arthritis, making it difficult to diagnose.<sup>1</sup> The most usual patient is a young woman of child-bearing age; however, this illness affects patients of all ages, ethnic backgrounds, and both sexes. Twenty per cent of all cases of lupus are diagnosed during the first two decades of life.<sup>2</sup>

The incidence is reported to be one per 1 000 in Caucasians, compared to four per 1 000 in African-Americans.<sup>1</sup> A review of rheumatic disorders in sub-Saharan Africa published in 2002, covering the period from the 1950s, found that SLE was increasing in the indigenous populations of East, Central and South Africa.<sup>3</sup> In most cases, the cause of SLE is unknown, although it is believed that many factors may be involved, including genetic predisposition and environmental factors, such as excessive sun exposure, infections, antibiotics (especially those in the sulpha and penicillin groups), extreme stress, certain drugs, and hormones.<sup>1</sup> It is known that, in families of patients with SLE, there is an increase in the number of relatives with SLE and rheumatoid arthritis compared with the normal population. Many of the relatives have abnormal proteins, such as antinuclear antibodies (ANAs), in their blood, although they may not have any symptoms of the disease.<sup>1,4</sup>

Some of the genes that increase a person's risk for SLE are known. For example, it has been found in the United States that a gene called DR2 increases a person's risk of developing lupus nephritis, although the vast majority of individuals with the gene are healthy.<sup>5</sup> There recently have been discoveries of a gene on chromosome 1 that is associated with lupus in certain families.<sup>5</sup>

Many researchers suspect that a special type of immune reaction causes the disease. It is believed that patients develop antibodies against their own tissues, as if they have been vaccinated against themselves.<sup>4</sup> These antibodies are known as auto-antibodies, and the type of allergy is called autoimmunity (or an allergy against oneself). Some people possess lupus "genes". Certain viruses, drugs, chemicals in the environment, or extreme emotional stress may activate the gene.<sup>5</sup> This gene encodes antibodies and/or other products that damage tissue, the net effect of which results in the surveillance system of the white blood cells (i.e. lymphocytes) ultimately stimulating the formation of antibodies.<sup>4,5</sup>

Although lupus can affect any part of the body, most people experience symptoms in only a few organs. Table I lists the most common symptoms of people with lupus.

A study of eight cases of large pericardial effusions due to SLE seen at Tygerberg Hospital reported that Raynaud's phenomenon, arthralgia and nephritis were the most common clinical features.<sup>6</sup> In a study of 36 cases of childhood SLE treated by the renal units of the Johannesburg and Chris Hani Baragwanath Hospitals, rashes, polyarthritis and renal pathology were the most common clinical features seen at diagnosis.<sup>7</sup>

**Table I:** Table of symptoms according to the Lupus Foundation of America<sup>4</sup>

Symptoms	Occurrence
Achy joints (arthralgia)	95%
Fever more than 100 degrees F (38 degrees C)	90%
Arthritis (swollen joints)	90%
Prolonged or extreme fatigue	81%
Skin rashes	74%
Anaemia	71%
Kidney involvement	50%
Pain in the chest on deep breathing (pleurisy)	45%
Butterfly-shaped rash across the cheeks and nose	42%
Sun or light sensitivity (photosensitivity)	30%
Hair loss	27%
Abnormal blood-clotting problems	20%
Raynaud's phenomenon (fingers turning white and/or blue in the cold)	17%
Seizures	15%
Mouth or nose ulcers	12%

**Table II:** Revised criteria for SLE of the American Rheumatism Association<sup>8</sup>

Features <sup>1</sup>	Characteristics
Malar rash	Fixed erythema, flat or raised, sparing the nasolabial folds
Discoid rash	Erythematous raised patches with adherent keratotic scarring and follicular plugging
Photosensitivity	Skin rash as a result of unusual reaction to sunlight
Oral ulcers	Oral or nasopharyngeal ulceration; may be painless
Arthritis	Non-erosive, involving two or more peripheral joints
Serositis	a. Pleuritis (convincing history of pleuritic pain, rub or pleural effusion) or b. Pericarditis (rub, ECG evidence or effusion)
Renal disorder	a. Persistent proteinuria >0.5 g/day, or b. Cellular casts (red cell, granular or tubular)
Neurological disorder	Seizures or psychosis in the absence of offending drugs or metabolic derangement
Haematological disorder	a. Haemolytic anaemia or b. Leucopenia <sup>2</sup> (<4000/mm <sup>3</sup> ) or c. Lymphopenia <sup>2</sup> (<1500/mm <sup>3</sup> ) or d. Thrombocytopenia <sup>2</sup> (<100 000/mm <sup>3</sup> ) in the absence of offending drugs
Immunology disorder	a. Anti-DNA antibodies in abnormal titre or b. Presence of antibody to Sm antigen or c. Positive antiphospholipid antibodies
Antinuclear antibody disorder	Abnormal titre of ANA by immunofluorescence

<sup>1</sup> For the purpose of identifying patients for clinical studies, a person must have SLE if any four out of 11 are present serially or simultaneously.  
<sup>2</sup> On two separate occasions.

Because many lupus symptoms mimic other illnesses, are sometimes vague and may come and go, lupus can be difficult to diagnose.<sup>4</sup> Diagnosis is usually made by a careful review of a person's entire medical history, coupled with an analysis of the results obtained in routine laboratory tests and some specialised tests related to immune status. Currently, there is no single laboratory test that can determine whether a person has lupus or not. To assist the physician in the diagnosis of lupus, the American College of Rheumatology (ACR) in 1982 issued a list of 11 symptoms or signs that help distinguish lupus from other diseases. This list has recently been revised.<sup>8</sup> A person should have four or more of these symptoms for lupus to be suspected (see Table II). The symptoms do not all have to occur at the same time.<sup>4</sup>

SLE is a complicated disease, as no patient presents with the same symptoms. This could be due to the surrounding environment, such as the climate, where sunlight plays a role in photosensitivity and skin rashes, or a colder climate could onset complications such as Raynaud's phenomenon. Central South Africa has a fluctuating climate, with extreme winters and summers. No study has been done in central South Africa (Free State) to describe the symptoms and complications of patients.

The aim of this study was to determine the most common symptoms and complications of patients with systemic lupus erythematosus (SLE) attending the SLE outpatient clinic at Universitas Hospital in Bloemfontein, South Africa.

### Methodology

The study population for this descriptive study, which was conducted in the second half of 2004, included all patients attending the SLE clinic at Universitas Hospital diagnosed with SLE according to the ACR classification criteria. Patients were only included if they had at least one follow-up visit. Patients with only discoid lupus were excluded.

Data were collected by means of an anonymous and confidential data sheet from the patients' files, which are stored at the SLE Clinic of Universitas Hospital. The researchers first went through the patients' files to ensure that the patients included in the study sample were known SLE patients. If the patient complied with the criteria, the required demographic and disease information were recorded.

A pilot study was conducted on the files of five SLE patients from the SLE Clinic at Universitas Hospital. The data sheet was amended after the pilot study.

Permission was obtained from the Ethics Committee, Faculty of Health Sciences, University of the Free State to conduct the study. The CEO of Universitas Hospital gave approval for the study to be conducted at the hospital and for the files of the relevant patients to be used.

### Results

Data were obtained from 76 patients: 71 females (94.7%) and five males (5.3%). The patients' ages at the initial visit ranged from 13.8 to 80.6 years, with a mean of 38.6 years. Fifty per cent of the patients were between the ages of 21 and 39. Eighteen per cent of the files did not indicate the patient's race. Of the patients for whom race was known, African patients accounted for 61.3%, whites for 33.9%, Asians for 1.6% and coloureds for 3.2%.

Figure 1 provides a summary of the systems involved in the body. Patients most frequently had immunological (90.8%), mucocutaneous (86.9%), musculoskeletal (85.5%) or cardiovascular problems (77.6%).

Table III provides a summary of the different types of immunological blood tests relating to SLE that were done. ANA was positive in 88.2% of the patients.

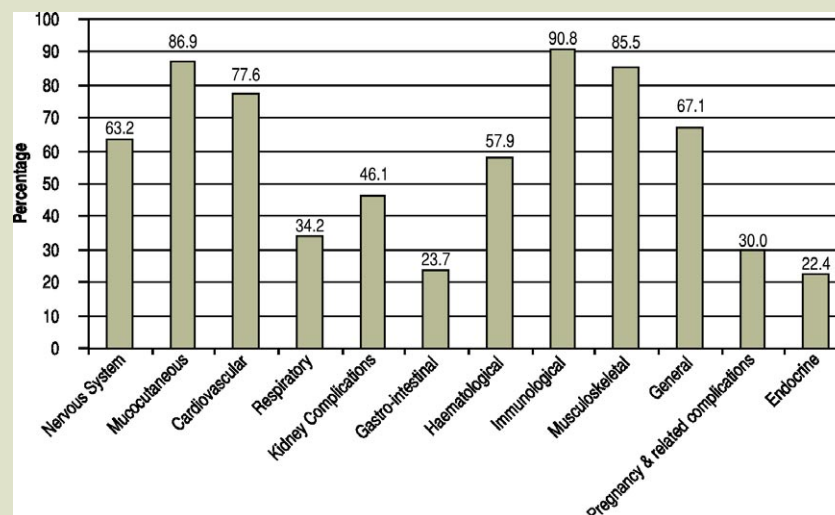
Malar skin was the most prevalent (69.7%) among the mucocutaneous problems, followed by photosensitivity (53.9%) and mouth ulcers (47.4%). In relation to the musculoskeletal system, patients mainly had aches (77.6%) and arthritis (55.3%). Cardiovascular problems were mainly hypertension (47.4%), Raynaud's phenomenon (30.3%) and vasculitis (19.7%).

Fatigue was the most common general symptom, occurring in 57.9% of patients, followed by general malaise in 29.0%.

**Table III:** Results of the immunological blood tests

Immunological blood tests	Percentage (%) positive
Antinuclear antibody (ANA)	88.2
Anti-DNA	40.8
Anti-Sm	42.1
Anti-RNP	48.7
Anti-SSA	46.1
Anti-SSB	32.9

**Figure 1:** Systems involved in SLE patients (n=76)



Regarding respiratory problems, 17.1% of the patients had pleuritis and 7.9% had tuberculosis.

### Discussion

SLE is far more prevalent in females. This could be due to hormonal factors and the fact that females of many mammalian species have higher antibody responses than males.<sup>9</sup> The findings of this study are consistent with the statistics of the Lupus Foundation of America, which found that nine out of 10 SLE patients are female.<sup>4</sup>

Diagnostically, the most important antibodies to detect are ANA, since this test is positive in more than 85% of patients, usually at the onset of symptoms. However, being ANA positive does not necessarily indicate SLE. Almost all patients with SLE are ANA positive in significant titres. The results of the ANA test have to be interpreted in the light of the patient's medical history, as well as the clinical signs and symptoms.

Musculoskeletal manifestations are also indicative of active disease. Most people with SLE have intermittent polyarthritis, varying from mild to disabling and characterised by soft tissue swelling and tenderness in the joints.<sup>9</sup> Athralgia may be present without arthritis. Myopathy may also be associated with polymyositis overlap, or may be a complication of chronic steroid use.


Headaches are common; when excruciating they often indicate SLE flare; when milder they are difficult to distinguish from migraine and tension headaches.<sup>9</sup>

The high prevalence of mucocutaneous manifestations may be due to the harsh climate of the Free State. This is evident in the high percentage of patients who have a malar rash. Worsening of this rash often accompanies a flare-up of systemic disease. Patients with mucocutaneous manifestations are extremely photosensitive, and most have antibodies to SSA.<sup>9</sup> Photosensitivity, along with mouth ulcers, is also indicative of a flare-up.

Within the cardiovascular system, Raynaud's phenomenon was more prevalent compared to the findings of the Lupus Foundation of America.<sup>4</sup> Once again, this could be due to the cold climate of the

Free State during the winter. Hypertension is a common problem in the normal population of South Africa and is not specific to SLE.

Pleuritis associated with serositis is also common in the South African population. Tuberculosis should be suspected in patients presenting with respiratory problems, as it is prevalent in SLE. It therefore is important to monitor the respiratory system by x-rays and lung function tests.

It is recommended that the data sheet be used routinely for the capture of patient information to form a basis for future prospective studies, substudies of the different systems, and to liaise with other clinics (dermatology, nephrology and haematology). 

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