Systemic Lupus Erythematosus  
— A Challenging Disease

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Lupus Erythematosus is a complex and challenging disease that affects the lives of many thousands of individuals and their families. It is multidimensional with physical, emotional and psychological aspects that require multidisciplinary and patient-centred treatment and support.

What is Lupus?
Lupus Erythematosus is an inflammatory autoimmune disease. Lupus means “wolf”. Erythematous means “redness”. The facial rash that usually accompanies lupus looked like the bite of a wolf, hence the name ‘Lupus Erythematosus’.

1. Discoid Lupus Erythematosus (DLE)
Characteristics
- Skin rashes, sun sensitivity are the main problems. Lesions are patchy, crusted, sharply defined skin plaques that may scar. Organ involvement is rare.

2. Subacute Cutaneous Lupus Erythematosus (SCLE)
Characteristics
- Skin rashes, sun sensitivity are the main problems.

3. Drug-induced Lupus
Characteristics
- Develops after the use of certain drugs. Pleuropericardial inflammation, fever, rash and arthritis and serologic changes often occur. Subsides after the offending drug is discontinued.

Drugs with proven association are (a) Procainamide (b) Hydroxychloroquine (c) Isoniazid (d) Methyldopa (e) Chlorpromazine

4. Neonatal Lupus
Characteristics
- Uncommon form affects newborn babies of women with lupus. Skin rashes and changes in blood cells occur and rarely affect normal cardiac pacing and may need a pacemaker.

5. Systemic Lupus Erythematosus (SLE)
Characteristics
- Chronic, inflammatory multisystem disorder of the immune system.

Who gets Lupus?
Usually develops in young women of childbearing age (15-45 years), but many men and children also develop Lupus.

Etiology
- Actual cause is unknown.
- Several likely possibilities are there in the etiology of Lupus.

(a) Immune system Dysfunction
- Formation of auto antibodies (anti nuclear antibodies) and immune complexes in higher concentrations in Lupus patients. The immune complexes may be deposited in various body tissues leading to the inflammation that results in tissue damage.

(b) Genetics
- The extremely high occurrence of lupus in identical twins and the increased prevalence of lupus among first and second degree relatives of lupus patients suggest a genetic component. If a mother has lupus, her daughter’s risk of developing the disease is 1:40 and her son’s risk is 1:250.

(c) Environmental influences
- Sunlight
- Exposure to sunlight can cause accumulation of certain cellular proteins, which react with auto antibodies and can lead to local or systemic inflammatory response.

(d) Stress
- Death of loved ones, divorce, job loss are some stress situations that can trigger lupus in susceptible individuals.

(e) Chemical substances in some medicines and (f) Viruses

(d) Hormones
- More prevalent in women in child bearing age, often flares during pregnancy and post partum period.

Symptoms of SLE
- Symptoms vary in individuals depending on the area involved. Systemic features include fever, anorexia, malaise, weight loss.

Cutaneous manifestation (Skin mucous membrane)
- Butterfly rash, periangual erythema, nail fold infarcts, alopecia, mucous membrane

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lesions in mouth or nose; Raynaud's phenomenon present in 20 per cent cases.

Musculo Skeletal
Arthritis often transient, seldom deforming morning stiffness, joint and muscle aching with or without synovitis.

Ocular
Conjunctivitis, Photophobia, transient blindness, blurring of vision, lupus rash on eyelids, ciliary bodies due to localized micro-infarction of the superficial nerve fiber layer on retina.

Cardio Pulmonary
Pleurisy, Pleural effusion, bronchopneumonia and pneumonitis, pericarditis, myocardits and valvular defects leading to hypertension and arrhythmias.

Gastro Intestinal
Abdominal pains and peritonitis due to vasculitis. Non-specific reactive hepatitis may alter liver function.
Psychosis, organic brain syndrome, seizures, peripheral and cranial neuropathies, transverse myelitis and strokes.

Renal
Several forms of glomerulonephritis. SLE kidney involve ment tends to remain silent until damage has occurred.

Haematologic
Anaemia, thrombocytopenia, leucopenia

Other clinical manifestations
Arterial and venous thrombosis, lymphadenopathy, spleno- megaly, Hashimoto's thyroiditis, repeated abortions, still births etc.

Diagnosis of SLE
Steps involved in the diagnosis of SLE
1. Complete medical history
2. Detailed physical examination
3. Performance of a whole battery of investigation. Complete blood count, ESR, renal and hepatic function tests, immunological tests for specific antibodies, etc are done. Abnormalities in these tests will lead to further tests like anti-id DNA antibody, anti-SM antibody, anti phospholipid antibodies. Lupus anti coagulant and anti cardiolipin antibody are phospholipids which are risk factor for venous and arterial thrombosis and foetal deaths.
4. Rule out other diseases like rheumatoid arthritis, osteoarthritis, mixed connective tissue disorder.
5. Close observation of the patient which may take months or years sometimes.

The American College of Rheumatology has developed a set of diagnostic criteria for SLE. Out of 11 criteria at least 4 should be satisfied to diagnose SLE

1. Malar rash (Butterfly rash)
2. Discoid rash
3. Photosensitivity
4. Oral ulcers
5. Arthritis
6. Pleuritis or pericarditis
7. Renal disorder
8. Neurological disorder
9. Haematological disorder
10. Immunological disorder
11. Abnormal ANA test

Management of patients with SLE
Although there is no cure for Lupus, a vast majority of patients can be treated successfully. Some mild form of SLE patients do not need any treatment although regular follow up is necessary. SLE patients who are on treatment can reasonably hope that a remission will occur when little or no treatment will be needed. Steps involved are:

(A) Physical rest
Fatigue persist despite normal blood reports. 8-10 hours of sleep at night with short periods of rest during the day is advisable. A well designed exercise programme to maintain strength, endurance and overall fitness. Patient and family can use fatigue as a guide to activity and when to stop for rest. Changes in lifestyle a pattern of daily living, change of occupation may be needed.

(B) Emotional rest
Disease itself as well as the side effects of corticosteroids can cause psychological problems in a lupus patient. Counseling for both patient and family. Emotional stressors of SLE patients should be carefully assessed. Meditation, Yoga and communication with friends and relatives and developing a positive attitude are some methods to relieve stress.

(C) Protection from Direct Sunlight
About 1/3 of lupus patients are photosensitive. All lupus patients should avoid direct prolonged exposure to the sun between 10 AM and 4 PM. Sensitive patients should apply a sunscreen with a sun protection factor (SPF) of at least 15. Avoid reflected lights, fluorescent and halogen lights.

(D) Diet and Nutrition
No specific diet recommended. Diet should be

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