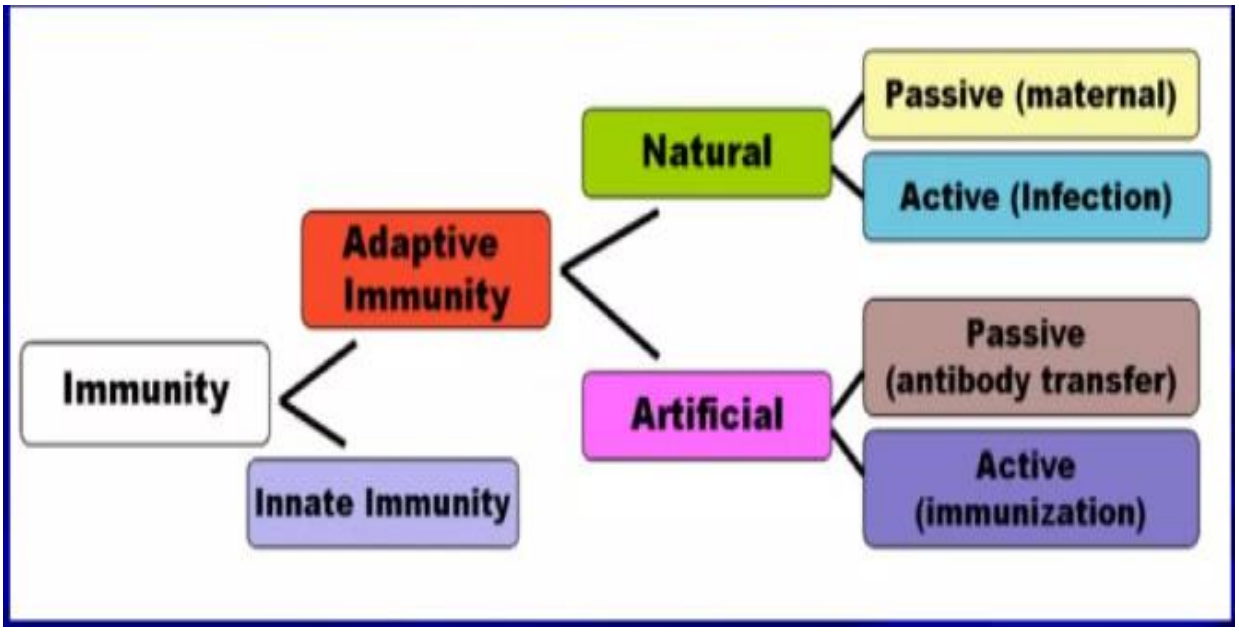
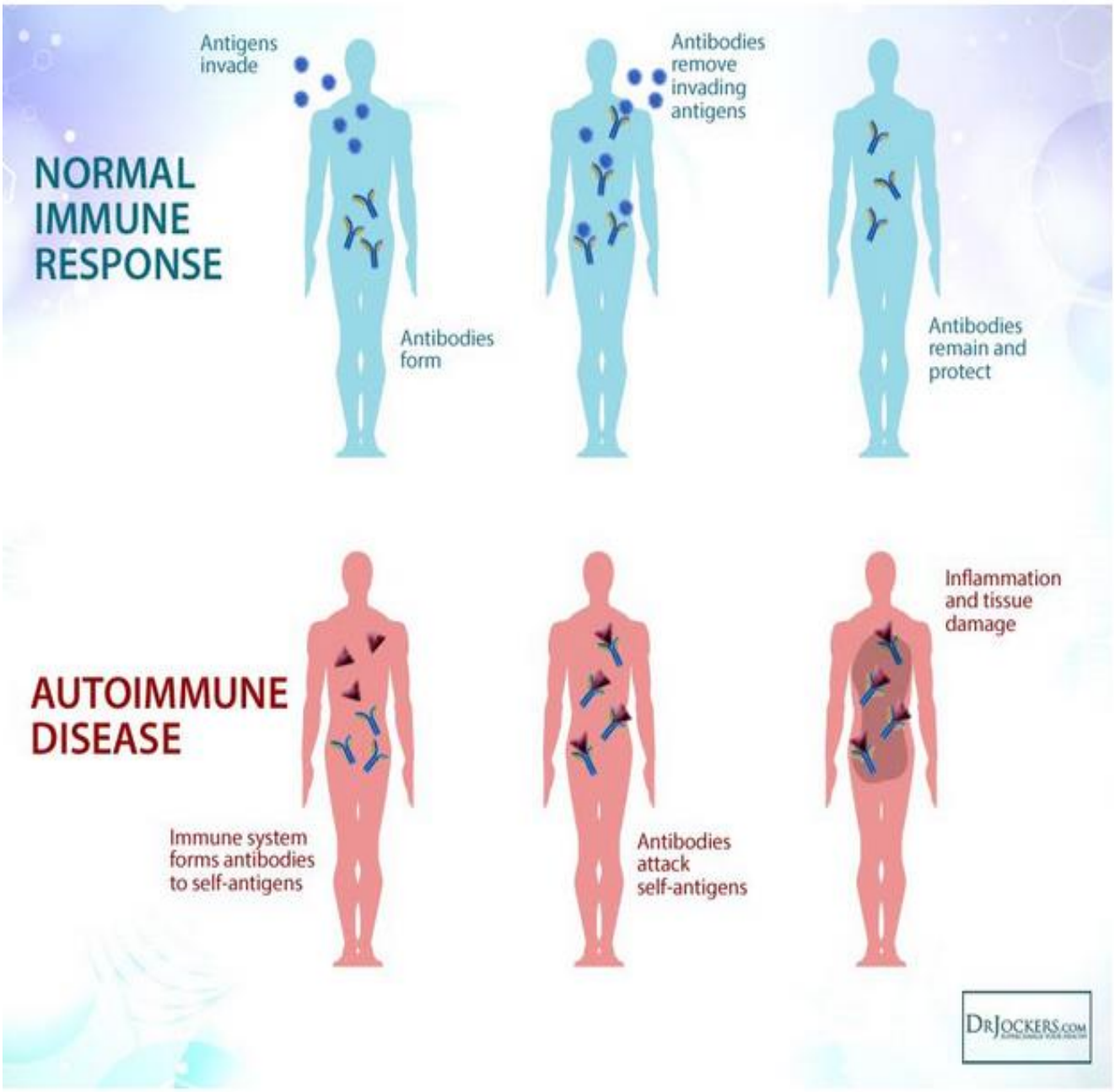
Autoimmune disorders

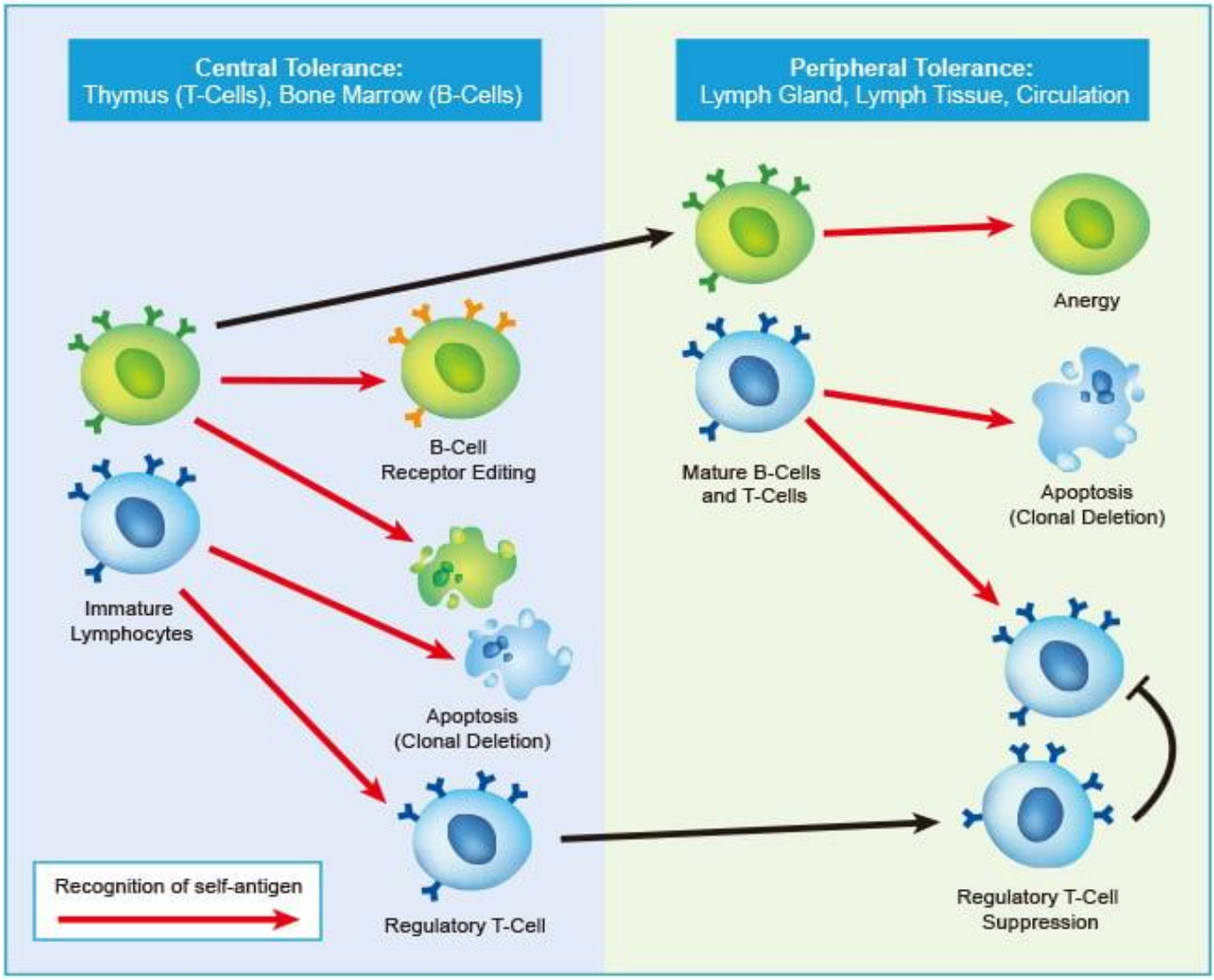
A healthy immune system defends the body against disease and infection. But if the immune system malfunctions, it mistakenly attacks healthy cells, tissues, and organs. Called autoimmune disease, these attacks can affect any part of the body, weakening bodily function and even turning life-threatening

A person's genes in combination with infections and other environmental exposures likely play a significant role in disease development.

Nearly 80% of people with a chronic autoimmune condition are women. These autoimmune diseases include rheumatoid arthritis, multiple sclerosis, scleroderma, lupus, Sjögren's syndrome




NORMAL IMMUNE RESPONSE
Antigens invade
Antibodies form
Antibodies remove invading antigens
Antibodies remain and protect
AUTOIMMUNE DISEASE
Immune system forms antibodies to self-antigens
Antibodies attack self-antigens
Inflammation and tissue damage
DrJockers.com



- In oral cavity auto antibodies are directed against the components of the epithelium, either against the cell attachments like desmosomes or cell to basement membrane attachment like hemi-desmosomes or against the basement membrane itself.

Autoimmune disorders fall into two general types:

1-Those that damage many organs (systemic autoimmune diseases)

2- Those where only a single organ or tissue is directly damaged by the autoimmune process (localized).

Systemic Autoimmune Diseases

- Rheumatoid arthritis (RA) and Juvenile RA (JRA) (joints; less commonly lung, skin
- Lupus [Systemic Lupus Erythematosus]) skin, joints, kidneys, heart, brain, red blood cells, other
- Scleroderma (skin, intestine, less commonly lung(
- Sjögren's syndrome (salivary glands ،tear glands, joints(
- Goodpasture's syndrome (lungs, kidneys)
- Wegener's granulomatosis (blood vessels, sinuses, lungs, kidneys(
- Polymyalgia Rheumatica (large muscle groups(
- Guillain-Barre syndrome (nervous system(

Localized Autoimmune Diseases

- Type 1 Diabetes Mellitus (pancreas islets)
- Hashimoto's thyroiditis, Graves 'disease (thyroid)
- Celiac disease, Crohn's disease ،Ulcerative colitis (GI tract)
- Multiple sclerosis , Addison's disease adrenal(
- Primary biliary cirrhosis, Autoimmune hepatitis (liver)
- Temporal Arteritis / Giant Cell Arteritis (arteries of the head and neck(

Systemic Lupus Erythematosus

Systemic lupus erythematosus (SLE) is a multisystem autoimmune inflammatory disorder of unknown etiology.

An important feature is the formation of antibodies to DNA, which are associated with lupus nephritis. In the United States, SLE has a prevalence of approximately 1 in 2000 with a woman to man ratio of 9:1

The prevalence is estimated to be two- to fourfold higher in non-Caucasian populations than in Caucasian populations.

In addition to systemic and isolated cutaneous lupus (chronic discoid lupus).

a distinct syndrome of drug-induced lupus is recognized. Unlike SLE, drug induced lupus rarely affects the kidney and is reversible on discontinuation of the offending agent.

Autoimmune diseases are more frequent in women than in men.

It is felt that the **estrogen** of females may influence the immune system to predispose some women to autoimmune diseases.

• An autoimmune inflammatory disorder of unknown etiology, primarily of young women, it can affect many parts of the body, including the joints, skin, kidneys, heart, lungs, blood vessels, and brain.

• In SLE, organ injury is secondary to either the direct binding of autoantibodies to self-antigens or the deposition of immunocomplex in vessels or tissues.

- An important feature is the formation of antibodies to DNA, which are associated with lupus nephritis

Lupus erythematosus either:-

1-systemic lupus erythematosus
2- Isolated cutaneous lupus (chronic discoid lupus)
3-drug-induced lupus is recognized.

Unlike SLE, drug-induced lupus rarely affects the kidney and is reversible on discontinuation of the offending agent (medication).

ETIOLOGY

Although the exact etiology of SLE is unknown, a complex interplay of genetic and environmental factors that leads to a progressive loss of peripheral tolerance and production of autoantibodies is believed to be crucial for SLE initiation.

1-Genetic Factors.(SLE in dizygotic twins is 3%, whereas it is up to 34% for monozygotic twins)

2- Environmental and Infectious Factors (viruses, vaccines, medications and ultraviolet).

Environmental factors, including infections particularly with EBV and other viruses

3- Exposure to pollutants, hormonal factors, ultraviolet light and smoking

4- Diet has been linked to the development of SLE.

5- In addition, over 80 drugs, hydralazine, isoniazide, and procainamide are associated with drug-induced lupus.

Common Symptoms of Lupus:-

Painful or swollen joints and muscle pain

Unexplained fever

Red rashes, most commonly on the face

Chest pain upon deep breathing

Unusual loss of hair

Pale or purple fingers or toes from cold or stress (Raynaud's phenomenon)

Sensitivity to the sun

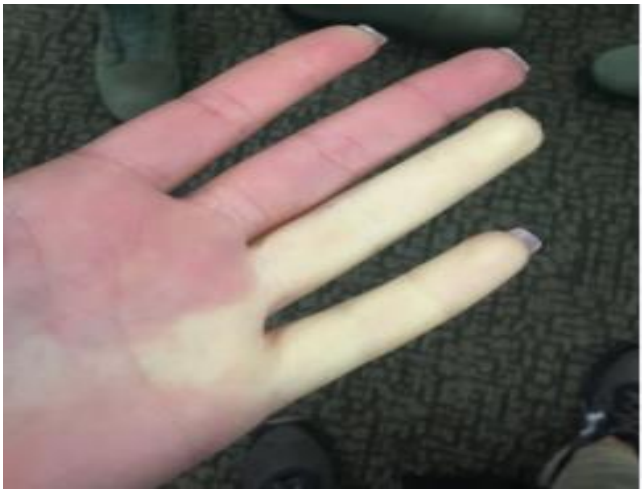
Swelling (edema) in legs or around eyes

Mouth ulcers

Swollen glands

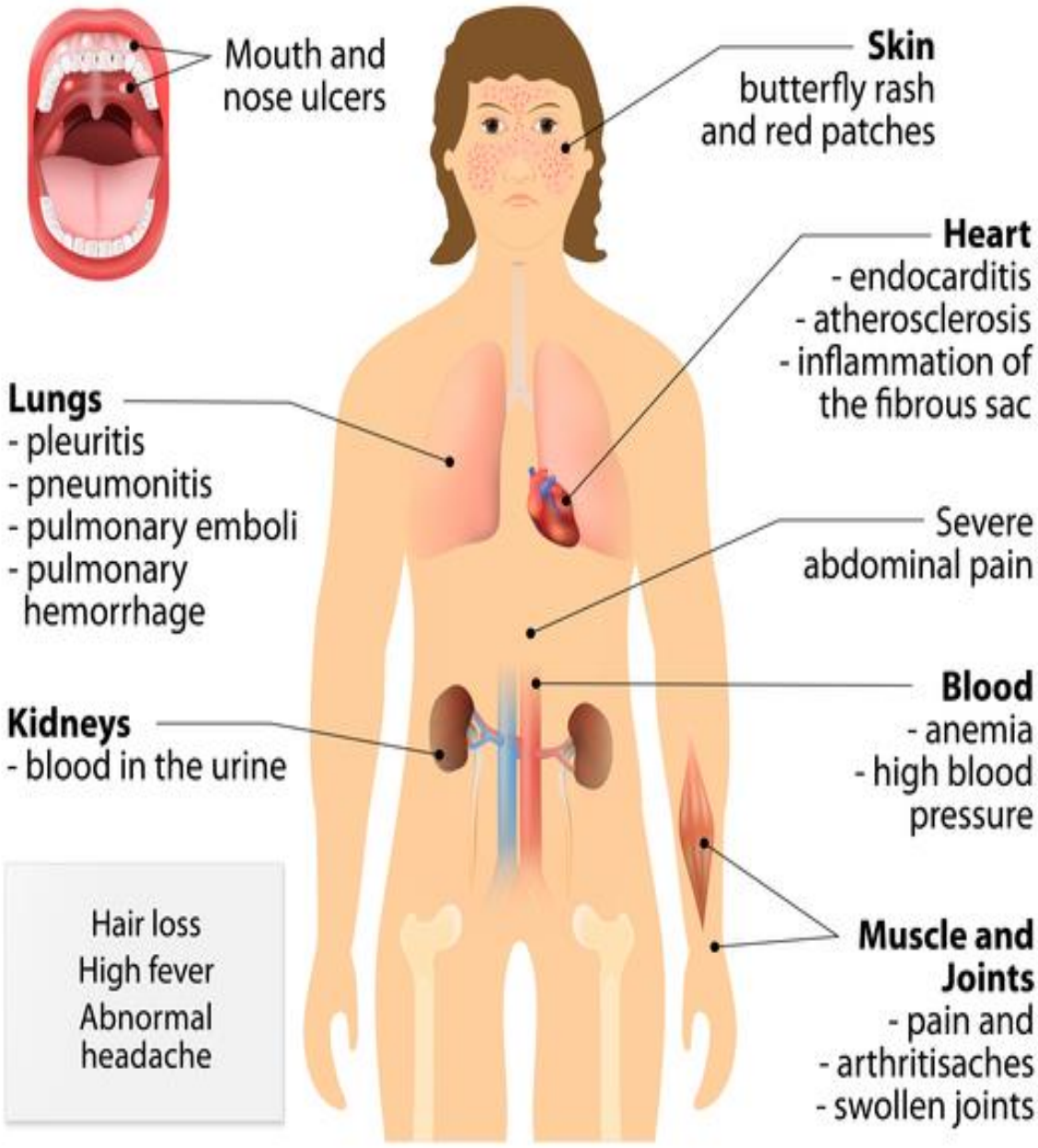
Extreme fatigue

Kidney problems



Raynaud's phenomenon

Systemic lupus erythematosus



Clinical Manifestations

Skin lesions of lupus can be classified into

1- lupus-specific (having diagnostic clinical or histopathological features)

2- Non specific lesions.

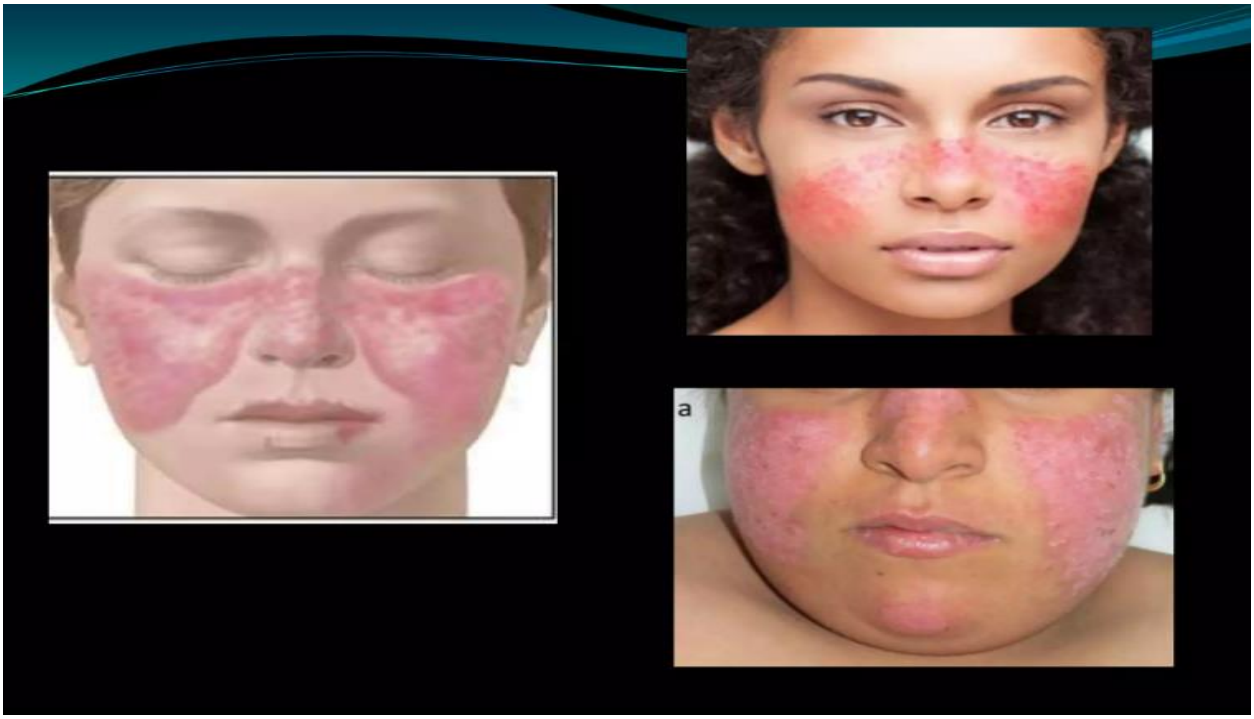
Three subtypes of lupus-specific skin lesions

a- Acute ,b- subacute, and c- chronic.

Acute cutaneous lupus occurs in 30%–50% of patients and is classically represented

by the butterfly rash—mask-shaped erythematous eruptions involving the malar areas and bridge of the nose.

Bullous lupus and localized erythematous papules also belong to the acute lupus category.

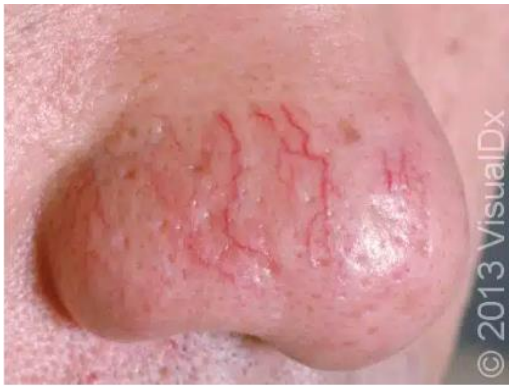


Chronic cutaneous lupus occurs in 15%–20% of cases and affects the skin of the face or scalp in about 80% of cases.

Non specific but suggestive skin manifestations of lupus are common and include alopecia (both scarring, following discoid lesions and non scarring), photosensitivity,

Raynaud's phenomenon, urticaria, erythema, telangiectasia,

and cutaneous vasculitis.



Oral Manifestations

1- Oral ulcerations,

2- Erythematous lesions,

3- Discoid lesions.

Estimates of prevalence vary from 9% to 45% in systemic disease and from 3% to 20% in localized cutaneous disease.

Oral ulcerations are listed among the criteria for SLE diagnosis .These ulcerations cannot be easily distinguished from other common oral conditions, such as aphthous ulcers,

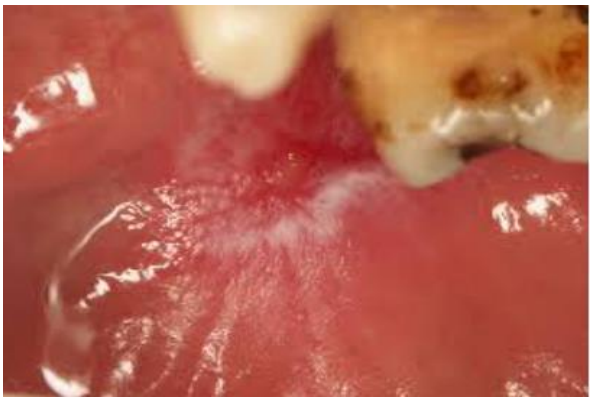
They occur with increased frequency on the palate and in the oropharynx and are characteristically painless

Discoid oral lesions are similar to those occurring on the skin and appear as whitish striae frequently radiating from the central erythematous area, giving a so-called brush border.

Atrophy and telangiectases are also frequently present.

Buccal mucosa, gingiva, and labial mucosa are the most commonly affected intraoral sites.

Isolated erythematous areas are also common, especially on the palate. It may be difficult to differentiate these lesions from other common mucosal disorders such as oral candidiasis or lichen planus, especially if there are few lesions and there is no systemic or cutaneous involvement.



The diagnosis of SLE, 11 criteria were established by the American Rheumatism Association.

1. malar (over the cheeks of the face) "butterfly" rash
2. discoid skin rash (patchy redness with hyperpigmentation and hypopigmentation that can cause scarring),
3. photosensitivity (skin rash in reaction to sunlight [ultraviolet light] exposure),
4. mucous membrane ulcers (spontaneous ulcers of the lining of the mouth, nose, or throat),
5. arthritis (two or more swollen, tender joints of the extremities),
6. pleuritis or pericarditis (inflammation of the lining tissue around the heart or lungs, usually associated with chest pain upon breathing or changes of body position),
7. kidney abnormalities (abnormal amounts of urine protein or clumps of cellular elements called casts detectable with a urinalysis),
8. brain irritation (manifested by seizures [convulsions] and/or psychosis),
9. blood-count abnormalities (low counts of white or red blood cells, or platelets, on routine blood testing),
10. immunologic disorder (abnormal immune tests include **anti-DNA or anti-Sm** [Smith] antibodies, falsely positive blood test for syphilis, anticardiolipin

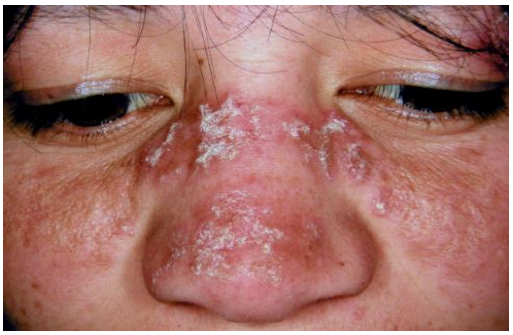
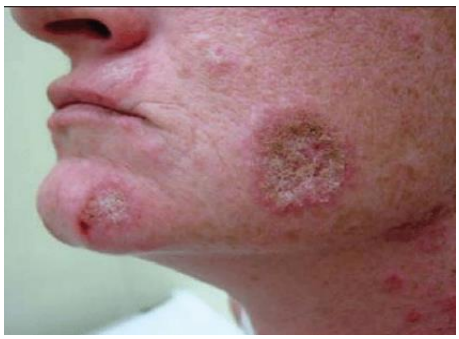
antibodies, lupus anticoagulant)

11.Antinuclear antibody (**positive ANA antibody testing)**

A person has SLE if any 4 out of 11criteria are present simultaneously or serially on two separate occasions.

Discoid lupus erythematosus

- DLE is essentially a skin disease with mucocutaneous lesions indistinguishable clinically from those of systemic lupus.
- Significant autoantibody production is present.
- **It occurs predominantly in females in the third or fourth decade of life.**
- Typical cutaneous lesion appear as red patches in sun-exposed area, such as face, extremities, these lesions **expand by peripheral extension and are usually disk-shaped**.

Signs

well-defined red plaques with an adherent scale and
follicular plugging which may result in scarring and post-inflammatory hyperpigmentation

Management of LE :-

Diagnosis:- of SLE should be by the pattern of antinuclear autoantibodies. The most specific is that to **double-stranded DNA.(ANA, Anti dsDNA)**

Hematological finding in active SLE (raised ESR, anemia and leukopenia or thrombocytopenia).

Treatment:- oral lesion of DLE may respond to topical corticosteroids. Oral lesions in acute SLE may not respond to dose of corticosteroids adequate to control systemic effect of the disease, palliative treatment is need until the disease activity is decrease.

Ttreatment of systemic lupus erythematosus

1- There is no permanent cure for SLE. The goal of treatment is to relieve symptoms
and protect organs by decreasing inflammation and/or the level of autoimmune activity in the body.

2- The precise treatment is decided on an individual basis. Many people with mild symptoms may need no treatment or only intermittent courses of anti-inflammatory medications.

3- Those with more serious illness involving damage to internal organ(s) may require
high doses of corticosteroids in combination with other medications that suppress the body's immune system.

Avoidance of sun exposure is very important

Nonsteroidal anti-inflammatory drugs (NSAIDs) are helpful in reducing inflammation and pain in muscles, joints, and other tissues.

Corticosteroids are more potent than NSAIDs in reducing inflammation and restoring function when the disease is active.

Corticosteroids are particularly helpful when internal organs are affected.

Hydroxychloroquine (Plaquenil) is an antimalarial medication found to be particularly effective for SLE people with fatigue, skin involvement, and joint disease. Consistently taking Plaquenil can prevent flare-ups of lupus.

Medications that suppress immunity (immunosuppressive medications) are also called cytotoxic drugs used for treating people with more severe manifestations of SLE, such as damage to internal organ(s).

Examples of immunosuppressive medications include methotrexate azathioprine (Imuran), cyclophosphamide (Cytoxan)

Dental Management

1- Risk of Infection

Daily treatment with higher doses of prednisone (over 7.5 –10 mg/day) or other glucocorticoids, treatment with high doses of cyclophosphamide, and high disease activity are risk factors for infection in SLE patients.

If possible, elective oral surgical procedures with the potential for bacteremia should be delayed until the absolute neutrophil count is over 1000 cells/mm3, as neutropenia may be transient and respond to treatment with glucocorticoids.

If an oral procedure cannot be postponed, prophylactic antibiotics can be considered.

2- Risk of Bleeding

Traditionally, platelet transfusions have been recommended in surgical patients with platelet counts below 50,000 per mm3.

For patients with lupus who are receiving anticoagulants, established guidelines should be followed.

In general, oral surgical procedures are safe in patients taking warfarin with

therapeutic international normalized ratio (INR) ranges (2 –3.5) and do not require discontinuation of anticoagulation.