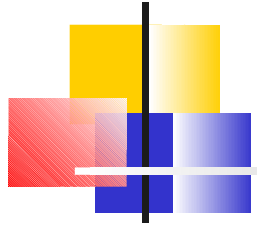


***SYSTEMIC LUPUS***

***ERYTHEMATOSUS***



Definition of SLE

Causes of SLE

Clinical Picture of SLE

Investigations of SLE

Treatment of SLE



# Definition

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- Systemic lupus erythematosus (SLE) is a multi-system auto-immune disease that is caused by tissue damage resulting from antibody and complement fixing immune complex deposition
- It is characterized by states of exacerbation and remission



## Definition (Cont.)

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- The immune system loses the ability to differentiate between foreign cells and it's own cells and tissues
- Antibodies against the immune system are formed
- The immune complexes that are formed build up in the tissue causing inflammation, injury to the tissue, and pain



# INCIDENCE OF LUPUS

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- Between 500,000 to 1.5 million Americans have lupus.
- 80-90% of lupus patients are female.
- 80% of lupus patients are between 15 and 45 years of age.
- Lupus affects more African Americans, Asian Americans, Hispanics, and Native Americans than Caucasians.



# WHAT CAUSES LUPUS?

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Genetics

Hormones

Environment



# Etiology

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- The specific cause is unknown
- Genetic factors may play a role
- Environmental agents
- Drugs or other chemical agents
- Dietary factors
- Ultraviolet radiation
- Infectious agents



# LUPUS IS...

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- Different for each person.
- A disease that ranges from mild to life threatening.
- Characterized by flares and remissions.





# TYPES OF LUPUS

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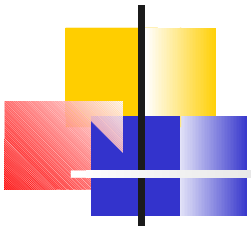
- Discoid or Cutaneous Lupus (DLE)
- Drug Induced Lupus (DIL)
- Neonatal Lupus
- Systemic Lupus Erythematosus (SLE)



# DISCOID LUPUS

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- Affects the skin, hair or mucous membranes.
- Identified by a rash or lesions.
- Diagnosed by biopsy of rash.
- 10% will evolve into SLE.
- Treatment includes topical or interlesional steroids; antimalarials.



# Discoid Rash





# DRUG INDUCED LUPUS

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- Develops after long-term use of certain medications.
- Most common in men over 50 years old.
- Symptoms are similar to SLE.
- Most important treatment is to recognize medication and discontinue use.
- Once medication is stopped, symptoms usually disappear completely within 6 months.



# NEONATAL LUPUS

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- Occurs when the mother's antibodies cross over the placenta to the baby.
- Can affect the skin, heart, liver and/or blood of the fetus and newborn.
- Good prenatal care can prevent most problems.



# SYSTEMIC LUPUS ERYTHEMATOSUS

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- Can affect any organ in the body including the joints, skin, lungs, heart, blood, kidney, or nervous system.
- Can range from mild to life threatening.
- No two people will have identical symptoms.



# COMMON SYMPTOMS OF SLE

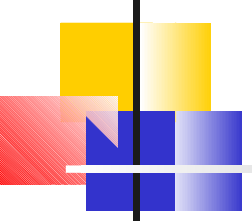
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- Achy joints
- Fever
- Fatigue
- Skin Rashes



# OTHER LUPUS SYMPTOMS

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- 
- Chest pain
  - Hair loss
  - Mouth sores
  - Photosensitivity
  - Anemia
  - Repeated miscarriages
  - Headache
  - Dizziness
  - Depression
  - Seizures
  - Memory disturbances

# ORGAN INVOLVEMENT WITH LUPUS

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- Kidneys
- Lungs
- Central nervous system
- Blood vessels
- Blood
- Heart



# DIAGNOSING LUPUS

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- Medical history (including family history)
- Complete physical examination
- Laboratory tests
- Skin or kidney biopsy



# ACR DIAGNOSTIC CRITERIA

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## **Skin criteria**

1. Butterfly rash
2. Discoid rash
3. Photosensitivity
4. Oral ulcers

## **Systemic criteria**

5. Arthritis
6. Serositis
7. Kidney disorder
8. Neurologic disorder

## **Laboratory criteria**

9. Hematologic abnormalities
10. Immunologic disorder
11. Antinuclear antibody

## New ACR and EULAR criteria for classification of SLE

All patients classified as having systemic lupus erythematosus must have a serum titer of antinuclear antibody of at least 1:80 on human epithelial-2-positive cells or an equivalent positive test. In addition, a patient must tally at least 10 points from these criteria. A criterion is not counted if it has a more likely explanation than SLE. Occurrence of the criterion only once is sufficient to tally the relevant points, and the time when a patient is positive for one criterion need not overlap with the time when the patient is positive for other criteria. SLE classification requires points from at least one clinical domain, and if a patient is positive for more than one criterion in a domain only the criterion with the highest point value counts:

Clinical domains	Points	Immunologic domains	Points
<b>Constitutional domain</b>		<b>Antiphospholipid antibody domain</b>	
Fever	2	Anticardiolipin IgG >40 GPL or anti-β2GP1 IgG >40 units or lupus anticoagulant	2
<b>Cutaneous domain</b>		<b>Complement proteins domain</b>	
Nonscarring alopecia	2	Low C3 or low C4	3
Oral ulcers	2	Low C3 and low C4	4
Subacute cutaneous or discoid lupus	4	<b>Highly specific antibodies domain</b>	
Acute cutaneous lupus	6	Anti-dsDNA antibody	6
<b>Arthritis domain</b>		Anti-Smith antibody	6
Synovitis in at least two joints or tenderness in at least two joints, and at least 30 min of morning stiffness	6		
<b>Neurologic domain</b>			
Delirium	2		
Psychosis	3		
Seizure	5		
<b>Serositis domain</b>			
Pleural or pericardial effusion	5		
Acute pericarditis	6		
<b>Hematologic domain</b>			
Leukopenia	3		
Thrombocytopenia	4		
Autoimmune hemolysis	4		
<b>Renal domain</b>			
Proteinuria >0.5g/24 hr	4		
Class II or V lupus nephritis	8		
Class III or IV lupus nephritis	10		

MDedge News

<b>Criterion</b>	<b>Definition</b>
Malar Rash	Rash over the cheeks
Discoid Rash	Red raised patches
Photosensitivity	Reaction to sunlight, resulting in the development of or increase in skin rash
Oral Ulcers	Ulcers in the nose or mouth, usually painless
Arthritis	Nonerosive arthritis involving two or more peripheral joints (arthritis in which the bones around the joints do not become destroyed)
Serositis	Pleuritis or pericarditis (inflammation of the lining of the lung or heart)
Renal Disorder	Excessive protein in the urine (greater than 0.5 gm/day or 3+ on test sticks) and/or cellular casts (abnormal elements the urine, derived from red and/or white cells and/or kidney tubule cells)

Criterion	Definition
Neurologic Disorder	Seizures (convulsions) and/or psychosis in the absence of drugs or metabolic disturbances which are known to cause such effects
Hematologic Disorder	Hemolytic anemia , leukopenia , lymphopenia or thrombocytopenia. The leukopenia and lymphopenia must be detected on two or more occasions. The thrombocytopenia must be detected in the absence of drugs known to induce it.
Antinuclear Antibody	Positive test for antinuclear antibodies (ANA) in the absence of drugs known to induce it.
Immunologic Disorder	Positive anti-double stranded anti-DNA test, positive anti-Sm test, positive antiphospholipid antibody such as anticardiolipin, or false positive syphilis test (VDRL).

*Adapted from: Tan, E.M., et. al. The 1982 Revised Criteria for the Classification of SLE. Arth Rheum 25: 1271-1277.*

# Malar Rash





# Oral Ulcers



 DOIA  
(c) University Erlangen,  
Department of Dermatology  
Phone: (+49) 9131 - 85 - 2727



# Systemic Lupus Erythematosus



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- **Head and Neck Manifestations**
  - Malar rash first sign in 50%
  - Erythematous maculopapular eruption after sun exposure
  - Oral ulceration



# Musculoskeletal System

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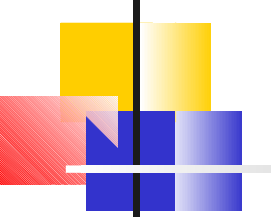
- All joints can be affected, however the wrists, knees, ankles, elbows, and shoulders are most common



# Musculoskeletal System (Cont.)

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- Hand deformities can include ulnar deviation and subluxation, swan neck deformities, and subluxation of thumb interphalangeal joints
- Reversible subluxation has been observed in the knees

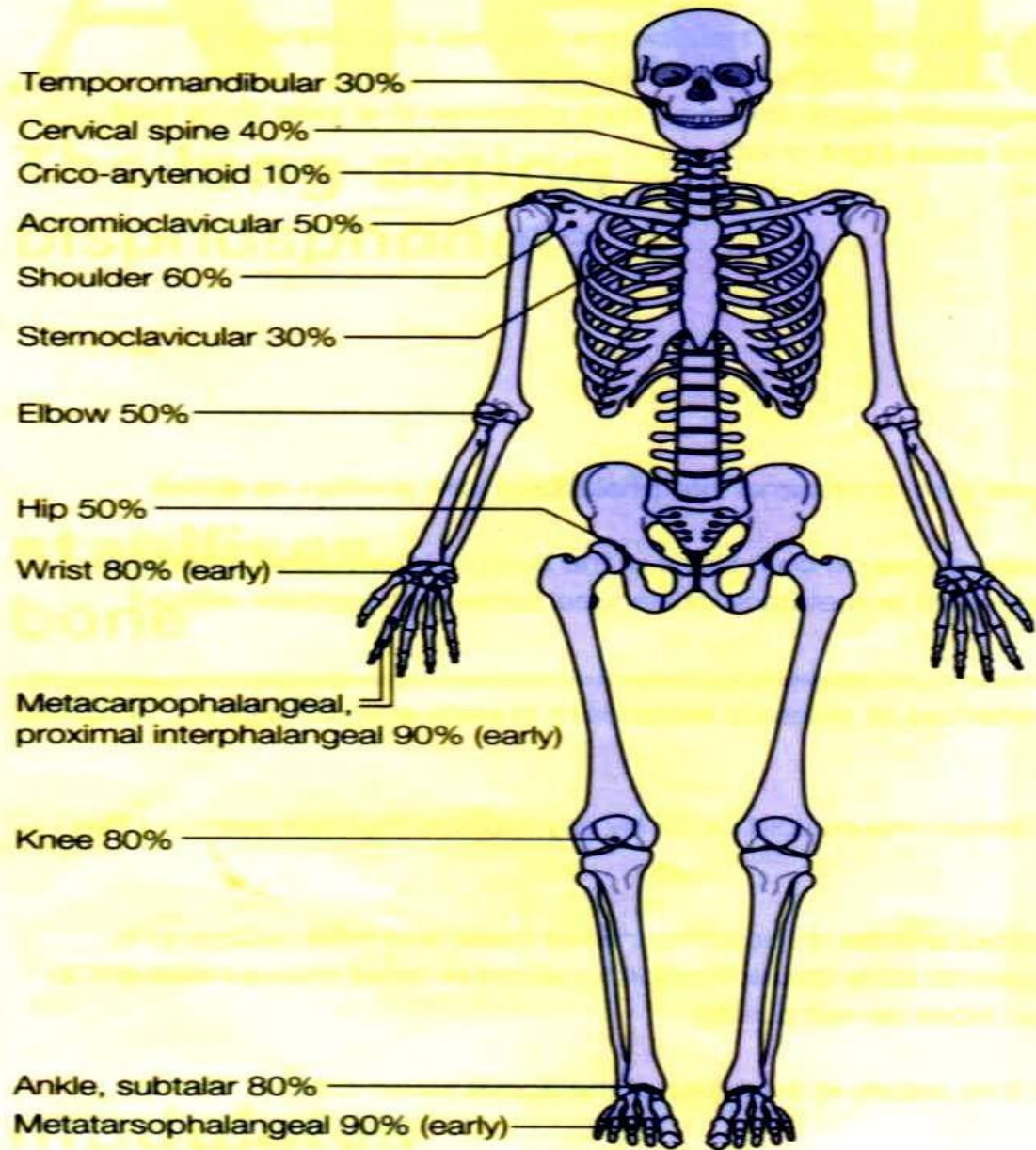


Boutonniere  
deformity  
of thumb

Ulnar deviation of  
metacarpophalangeal  
joints

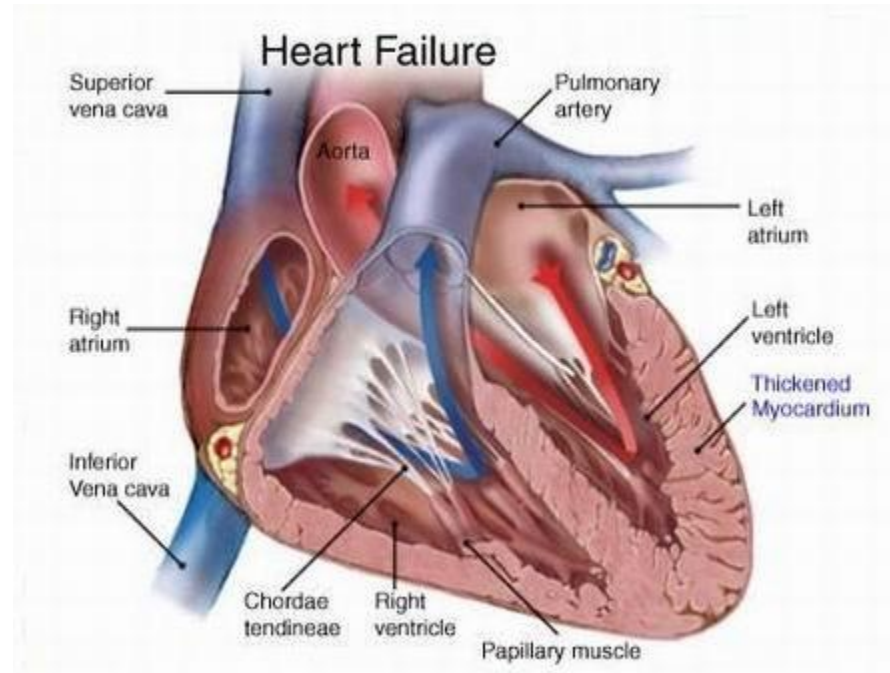
Swan-neck deformity  
of fingers

## Involvement of joint sites in established rheumatoid arthritis



# Cardiovascular System

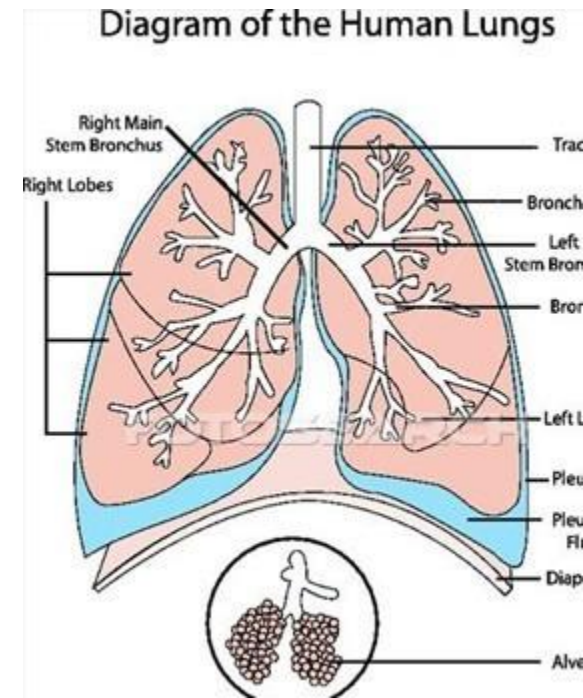
- Pericarditis is the most common cardiac manifestation
- myocarditis
- Endocarditis with characteristic lesion of the cardiac valve





# Pulmonary System

- Pleurisy is the most common manifestation of pulmonary involvement
- Interstitial lung disease
- Pulmonary embolisms





# Gastrointestinal Sytem

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- Difficulty swallowing
- Gastric reflux disease
- Anorexia, nausea, or vomiting may be present
- Pancreatitis

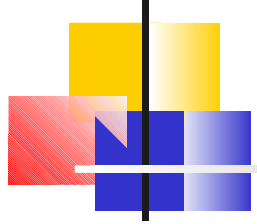
# Raynaud's Phenomena



[Http://www.dermis.net/doi](http://www.dermis.net/doi)

a

# Renal manifestations



- Tend to appear within the 1<sup>st</sup> 2yrs of SLE
- Almost 1/2 have asymptomatic urine abnormalities
  - Proteinuria - dominant feature
  - Haematuria – almost always present but not in isolation



# Nervous System

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- Neuropsychiatric manifestations of lupus occur frequently
  - May be mild to severe
  - Any location in the nerve system may be affected (brain, spinal cord, peripheral system)



# Nervous System (Cont.)

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- Central Nerve System
  - Acute confusional state
  - Psychosis
  - Anxiety disorder
  - Headache
  - Cerebrovascular disease



# Nervous System (Cont.)

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- Peripheral Nerve System
  - Cranial neuropathy
  - Mononeuropathy
  - Polyneuropathy

# COMMON LABORATORY TESTS

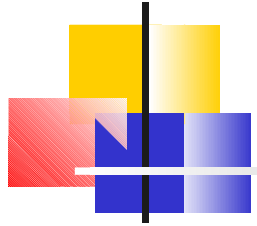
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- Antinuclear Antibody (ANA)
- Anti DNA
- Anti-Sm
- Anti-RNP
- Anti-Ro
- Anti-La



# OTHER LABORATORY TESTS

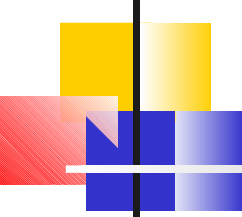
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- CBC (RBC, WBC, platelets)
- Urinalysis
- Sedimentation Rate (ESR)
- Rheumatoid Factor
- Skin biopsy
- Kidney Biopsy

# EFFECT OF LABORATORY TESTS WITH INCREASED LUPUS ACTIVITY

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- 
- C reactive protein (CRP)
  - Sedimentation rate (ESR)
  - Anti DNA
  - Liver and Kidney  
Function tests
  - CPK
  - Urine protein or cell casts
  - CBC (WBC, RBC,  
platelets)
  - Complement
  - Serum albumin



# LUPUS TREATMENT

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- Team effort
- Tailored
- Tentative



# LUPUS PHYSICIANS

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- Family Practitioner
- Internist
- Rheumatologist
- Clinical Immunologist
- Dermatologist
- Nephrologist
- Other specialists



# COMMON LUPUS MEDICATIONS

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- NSAIDs
- Antimalarials
- Corticosteroids
- Immunosuppressants
- Investigational (research)



# NSAIDs

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- Aspirin
- Ibuprofen
- Indomethacin



# CORTICOSTEROIDS

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- Prednisone
- Prednisolone
- Methylprednisolone
- Hydrocortisone



# ANTIMALARIALS

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- Hydroxychloroquine





# IMMUNE SUPPRESSANTS

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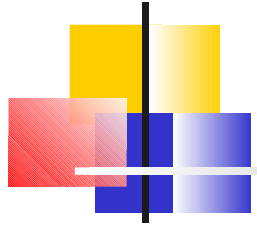
- Cyclophosphamide
- Mycophenolate mofetil  
/Azathioprine



# PREVENTIVE MEASURES

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- Sun precautions
- Rest
- Nutrition/diet
- Exercise
- Moist heat
- Prevent infection
- Don't smoke



*Thank You*

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