

# Systemic Lupus Erythematosis

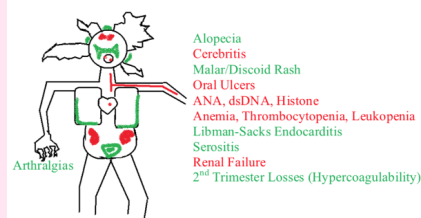
SLE has protean clinical manifestations that can affect virtually every organ, and can vary dramatically from patient to patient.  
 PRESENTATION: Active SLE, mild SLE, Uncontrolled SLE, Remission, Complete Response  
 NEED 4+ Manifestations as required by ACR

## SLE = ACR Diagnostic Criteria

- Malar Rash
- Discoid Rash
- Photosensitivity
- Oral Ulcers
- Arthritis
- Serositis OR
- Renal D/O OR
- Nero D/O OR
- Hematology D/O OR
- Immunologic D/O OR
- ANA Antibody

## SLE Management

- **Referral to specialist**
- General treatment considerations
- **SUN PROTECTION**
- Avoid Intense sunlight during peak daylight hours
- Apply sun screen **SPF 30** (30-60) prior to exposure; **reapply every 4 - 6 hours**
- Wear sun protective clothing
- **STOP smoking** makes skin condition worse
- Diet and nutrition
- Regular exercise
- Immunizations
- Avoid certain medications
- **No pregnancy during active disease**
- Avoid high dose **estrogen contraceptives**



## Topicals

- **Face Hydrocortisone - limit to 2 weeks**
- **Trunk/Limbs- Betamethasone Valerate**

## Systemic

- **Antimalarial - Hydroxychloroquine (Plaquinil)**
- **200-400 mg/day -NOT OVER 6.5 MG /KG/DAY**
- **SE:** GI upset, HA, Rash
- **Rare ADV- Retinal deposition with possible irreversible retinopathy**
- **NEEDS baseline DILATED EYE EXAM & yearly f/u**

## IF unable to use HYDRoxy use:

- **Chloroquine** (Aralin) **200-500 mg**
- **Quinacrine** 100 mg (does not cause renal toxicity but rarely used BC **YELLOW discoloration**)

## LUPUS with MSK S/S treatment:

- **NSAIDS**
- And/or **ACETAMINOPHEN**
- **PPI** may be needed
- **Methotrexate 7.5 - 15 mg/ day**
- **Initial:** Hydroxychloroquine
- **Second** Methotrexate
- **STEROIDS** - USE FOR FLARES
- **NSAIDS** - Arthralgia
- **Cyclophos** - Acute cerebrates/nephritis
- **Mycophenolate** - Maintenance
- **EYE EXAM** with **Hydroxychloroquine**
- No Eye S/S use **Quinacrine**

Pathogenesis is related in large part to production of increased quantities and immunogenic forms of nucleic acids and other self-antigens, which drive autoimmune-inducing activation of innate immunity, autoantibodies, and T cells.

AUTOIMMUNE CHRONIC  
 INFLAMMATORY DISORDER-  
 ETIOLOGY UNKNOWN

65% - between ages 16 & 55

20% - < 16

15% - > 55

In women of **child bearing age**, frequently **miscarriages** should be highly suspicious and one needs to consider the possibility of SLE, endocarditis, myocarditis, interstitial PNA, or aseptic meningitis's

## \*\*Variable;

**Most Common:** Picture of constitutional complaints with:

- **Skin, MSK, Mild hematologic and serologic**

## Some with:

- Hematologic, renal, or CNS involvement

**FEVER:** 50-100% of persons sometime during the disease presentation

Myalgia - common

**Loss** - prior to diagnosis of SLE; unintentional - decreased appetite, side effects of medications, GI (reflux, abdominal pain, PUD, pancreatitis)

## SLE- OLDER PERSONS LOL

- **S/S > 55 YO.** (10% of cases)
- Average age of onset 2- 3 yrs
- **F: M Ratio 4:1**
- Clinical Presentation
- Milder dais, less likely alopecia, malaria rash, photo sensitivity, oral/mucosal ulcers, glomerulonephritis, lymphadenopathy
- More likely: **CYTOPENIA, SEROSITIS, INTERSTITIAL PNA**
- Incidence of CA less except for **NON-HODKINGS LYMPHOMA**
- **Death- usually r/t complications o tx; INFECTIONS, CV DISEASE, STROKE**

**Management:** Varies with involved organ systems  
**Antimalarials** - well tolerated

**QUINACRINE** - Dose not cause **RETINAL** damage

**NSAIDS** - Use Cautiously with renal impairment

**Cyclophosphamide** - well tolerated

## Differentials:

- Chronic Fatigue
- Weight Loss
- Polyarthrits/polyarthralgia
- Glomerulonephritis; other renal
- Possible causes of abdominal pain, lung disease, cardiac, vision, neurologic, hematologic, lymphadenopathy, seizures, different skin lesions

**Prognosis** - Much improved - earlier diagnosis; better treatment

**Clinical Course variable** with remissions and acute and chronic relapses. The patter that **DOMINATES** during early disease tends to prevail.

## Prognostic factors:

Renal disease, HTN, **Male** gender, Young age, older age at new onset, **Presence of antiphospholipid** antibodies, **Poor socioeconomic status**, high overall disease activity

~ **90% of cases are in females; more in urban setting**

**Prevalence: Asian, AA, AfroCaribbeans and Hispanic Americans**

## UNDERLYING FACTORS

- **VIRUSES**
- **UV LIGHT**
- **SILICA DUST**
- **ENVIRONMENTAL:** Prevalence of lupus in dogs of pts with lupus; NO association between SLE and hair dyes, occupational solvent, pesticides or alcohol, NOTE: Moderate **alcohol intake** may be protective

**Gain-** Usually d/t salt and water retention associated with hypoalbuminemia (nephrotic syndrome or protein losing enteropathy or increased appetite associated with the use of glucocorticoids)

## GI Organ Involvement 25-40%

- **Dysphagia MOST FREQUENT**
- **Pulmonary** - MOST PATIENTS - **Pleuritic chest pain**
- **MSK**
- **Pleuritis**
- Upper Respiratory infection
- Interstitial lung disease 9%
- Pulmonary hypertension is **RARE**

## Neurologic and psychiatric 10-60% involvement

**MOST COMMON: \*\*\*Cognitive dysfunction; stroke; seizures; HA; peripheral neuropathy**

**LESS COMMON:** Movement D/O; Cranial neuropathies

Ocular involvement

Keratoconjunctivitis Sicca (Sjorgrens dry eye)

## Raynaud Phenomenon 16-40%

Secondary Raynaud's Phenomenon; Some type of underlying vascular dysfunction causing disordered vascular reactivity

**DRUG INDUCED-** 15K - 30K cases per year (equal in males and female but more common in older PT and Caucasians)

Possible Genetic predisposition:

**DRUGS: Procainamide, Hydralazine, Penicillamine, MONOCYCLINE** (YOUNGER patients) Su

Suspect - on medication **at least 1 month** and present with **TYPICAL** symptoms

**MANAGEMENT - STOP MEDICATION**

## • SLE Diagnostics: LABS:

- CBC and diff
- **CMP;** ESR and or CRP
- **CK**
- **UA**
- **Auto ANTIBODIES:**
  - ANA
  - Antiphospholipid antibodies
  - Antibodies to double stranded DNA
  - **Anti-SMITH (5m) Antibodies**
  - Serum Complement - C3 and C 4

## S/S CLINICAL PRESENTATION:

- **Fever, Fatigue, Weight loss,**
- **Photosensitivity Rash,**
- Arthralgia OR arthritis
- Raynaud Phenomenon
- **Serositis** (refers to inflammation of the serous tissues of the body, the tissues lining the lungs (pleura), heart (pericardium), and the inner lining of the abdomen (peritoneum) and organs within)
- Nephritis or Nephrotic Syndrome
- Neurologic Symptoms
- Alopecia
- Phlebitis
- **Recurrent Miscarriages**
- **Anemia**

## ORGAN INVOLVEMENTS:

- **MSK- 95%**
- Arthritis and arthralgia
- Migratory, symmetrical polyarticular
- **MORNING STIFFNESS - BRIEF**

## Musculocutaneous - Photosensitive lesions 80%

- **BUTTERFLY RASH**
- **SUBACUTE** cutaneous lesions
- Discoid lesions in malar distribution
- Alopecia
- Oral and or Nasal Ulcers (Purpura mucous membranes)
- **CARDIOVASCULAR** - Seen in most common presentation

- **Pericarditis**
- **Valvular Disease**
- **Myocarditis**
- **Increased risk of CAD**

## Renal ORGAN Involvement

**COMMON: Lupus Nephritis** with different classifications 1 - 6

**Other Forms:** Tubule-interstitial nephritis; vascular disease and renal disease associated with **drug-induced SLE**

## Hematologic:

**Cytopenia** (leukopenia, mild anemia of chronic disease and thrombocytopenia)  
**Thrombophilia** ( venous thromboembolism; arterial disease)  
 Lymphadenopathy and splenomegaly

## Immunologic:

- **Auto-antibodies - HALLMARK**
- **ANA + Titer**
- **Anti-double stranded DNA (dsDNA)**
- **Anti-SM antibodies**

## Imaging:

- Plain Radiograph of involved joints
- Renal US
- Chest X ray
- Echo
- EKG
- CT
- MRI
- Contrast Angiography
- Biopsy- tissue or organ

Treatment	When
Hydroxychloroquine	Everyone
Steroids	Flares only
IV Cyclophosphamide	Life-threatening illness
Mycophenolate Mofetil	Cerebritis, Nephritis
	Oral Cyclophosphamide
	Use after IV
Methotrexate	2 <sup>nd</sup> line to Hydroxychloroquine
NSAIDS	Symptom control only
Azathioprine	Adjunctive
Biologics	Maybe, not yet approved