



SYSTEMIC LUPUS

**SYSTEMIC INFLAMMATORY AUTOIMMUNE
CONDITION**

SYSTEMIC LUPUS

- ▶ **SYSTEMIC**

 - All bodily systems can be involved.

- ▶ **INFLAMMATORY**

 - Migration and infiltration of inflammatory cells into affected organs.

- ▶ **AUTOIMMUNE**

 - Failure of immune response to recognize self.

 - Over 80 different autoimmune conditions

 - Affecting 23.5 million Americans

SYSTEMIC LUPUS

A systemic illness that affects a multitude of organs and tissues within the body, individually or concurrently, with various degrees of severity.

The Impact of Lupus on the Body

Central and Peripheral Nervous System

Seizures, Psychosis, Headaches, Cognitive Dysfunction, Neuropathies, Depression, Low Grade Fever

Heart, Lungs

Pericarditis, Myocarditis, Endocarditis, Pleuritis, Pneumonitis

Kidneys

Edema, Hypertension, Proteinuria, Cell Casts, Renal Failure

Reproductive System

Pregnancy Complications, Miscarriages, Menstrual Cycle Irregularities

Blood

Anemia, Thrombocytopenia, Leukopenia, Thrombosis, Circulating Autoantibodies and Immune Complexes

Eyes and Mucous Membranes

Ulcers in the Eyes, Nose, Mouth or Vagina, Sjögren's Syndrome

Gastrointestinal

Nausea, Vomiting, Diarrhea, Weight Changes

Musculoskeletal

Extreme Fatigue, Arthralgia, Myalgia, Arthritis, Myositis

Skin

Butterfly Rash, Cutaneous Lesions, Photosensitivity, Alopecia, Vasculitis, Raynaud's Phenomenon



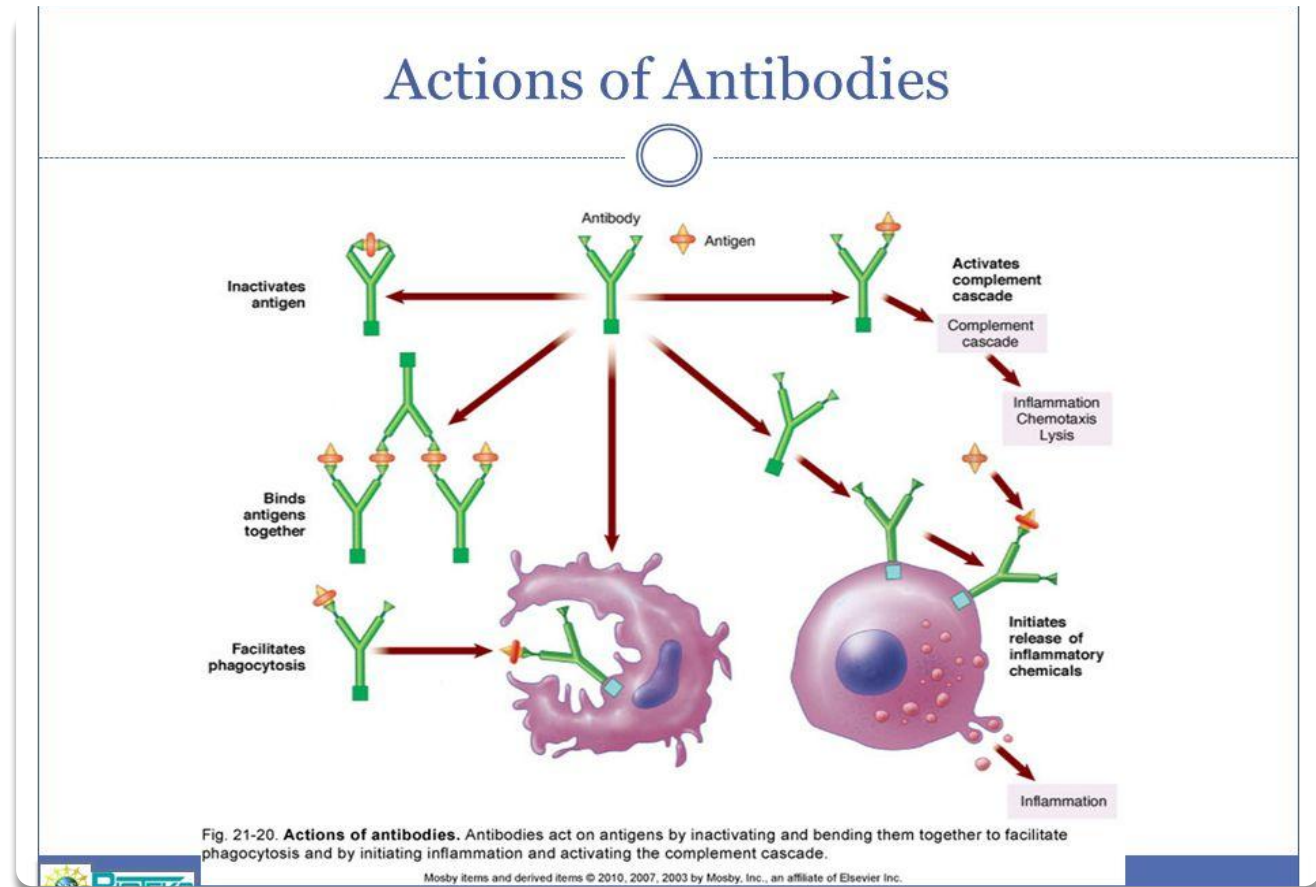
Lupus can affect any part of the body; however, most people experience symptoms in only a few organs.

- Lupus is an incurable chronic autoimmune disease that causes inflammation in various parts of the body. The disease can range from mild to life-threatening.
- The cause of lupus is unknown. Scientists believe that individuals are genetically predisposed to lupus, and that environmental factors "trigger" the symptoms.
- 90% of people with lupus are women, 80% of them developed lupus between ages 15 and 45.
- With proper treatment, most people with lupus can live a normal life span.



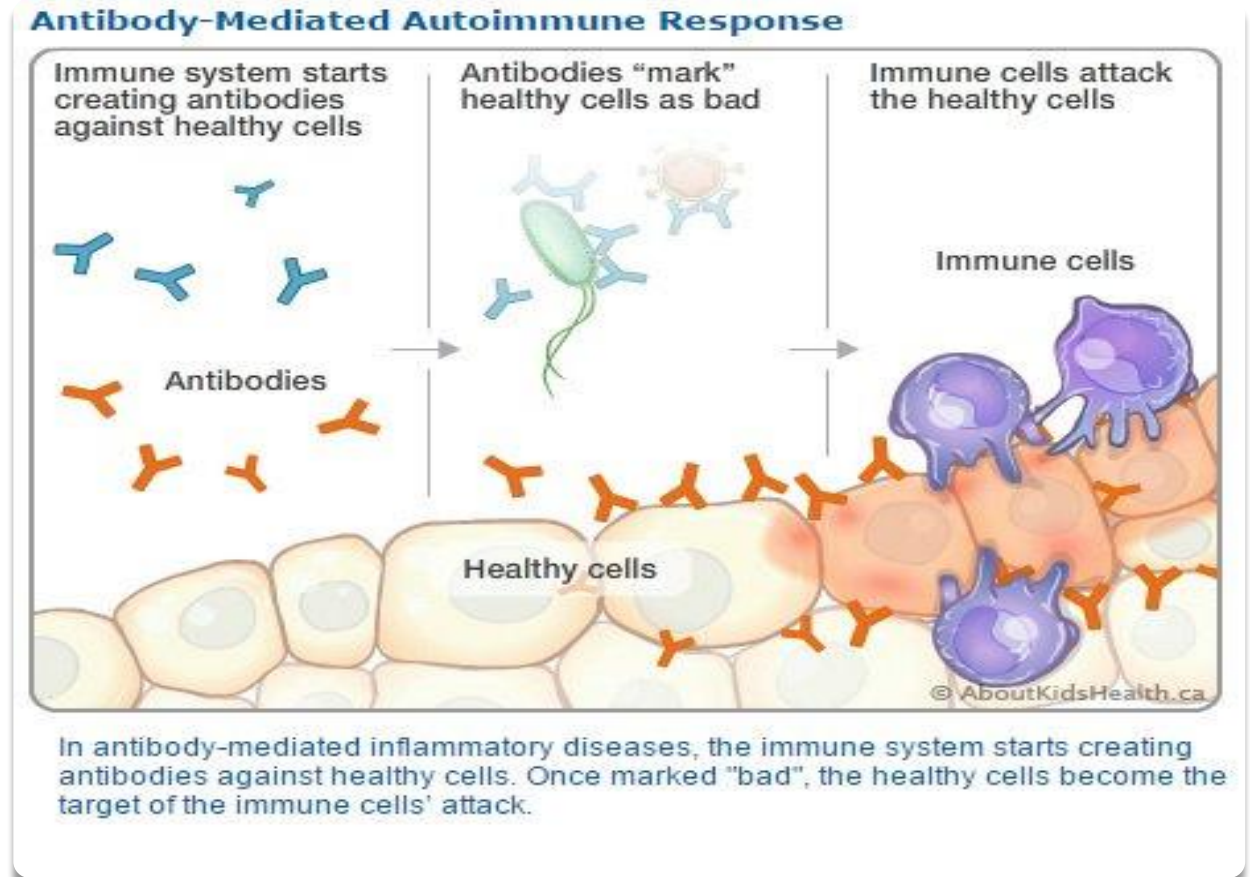
SYSTEMIC LUPUS

- ▶ Not only a diffuse systemic illness but is characterized by acute and chronic inflammation in the affected organ. And you can't have inflammation without this:



SYSTEMIC LUPUS

- ▶ Systemic lupus is not only a disease of multiple organs, and a disease of inflammation, it is most importantly a disease of “immunity gone bad” -
- ▶ AN AUTOIMMUNE DISEASE!



SYSTEMIC LUPUS

- ▶ About 1.5 million Americans have systemic lupus
- ▶ About 5-6 million people have the disease worldwide.
- ▶ Generally affects people between the ages of 15 – 45 years of age.
- ▶ More common in minority populations – African American, Asian, Native Americans, Native Hawaiians and Alaskans, and Pacific Islander's.
- ▶ African Americans “may” have the disease more frequently than Native Africans.
- ▶ 10 -15% of patients will die prematurely due to complications of the disease.

WHO GETS LUPUS?

African American
Women



Women of various
ethnic groups



Women of any age and
during pregnancy



Variable Nature of Lupus and Frequency of Presentation

- ▶ Constitutional symptoms (80-100%) - fatigue, myalgias, fever, weight loss.
- ▶ Arthritis/Arthralgias (65-90%) – often the earliest sign, one joint or many, migratory, symmetric, generally NOT deforming, 10% identical to RA.
- ▶ Dermatological (70-80%) – malar rash, photosensitivity, Discoid rash, alopecia (scarring, non scarring, patchy or diffuse), livedo reticularis, panniculitis.
- ▶ Renal (50%) – microscopic blood and protein loss in urine, most common internal organ involve, 2nd most common cause of death, hypertension, edema, 5 classes of SLE biopsy proven renal disease.
- ▶ Neurologic (25-75%) – headache, neuropsychiatric, seizures, strokes, myelitis, TIA, frequently associated with presence of antiphospholipids (APL), neuropsychiatric episodes.

Variable Nature of Lupus and Frequency of Presentation

- ▶ Pulmonary (14-100%) - due the large variable forms of involvement; pleurisy, pneumonitis, pulmonary HTN, pulmonary embolism, pulmonary fibrosis, alveolar hemorrhage.
- ▶ Gastrointestinal (50%) – oral ulcers, peritonitis, pancreatitis, bowel infarct, mesenteric vasculitis, autoimmune hepatitis, Lupoid hepatitis,
- ▶ Cardiac (33-40% {@ autopsy/echo}) – pericarditis, myocarditis, coronary vasculitis, Libman-Sacks endocarditis, accelerated CAD due to plaque.
- ▶ HOST OF OTHERS: Ocular, vascular, obstetric (3% of babies born have neonatal lupus) , endocrine (24% have thyroid disease), hematologic

HOW TO MAKE THE DIAGNOSIS OF LUPUS

YOU MUST HAVE 4 OF THE 11
CRITERIA.

NO ONE CRITERIA MAKES
THE DIAGNOSIS OF
LUPUS!!!

- ▶ 1. Malar rash – a butterfly shaped rash across cheeks and nose
- ▶ 2. Skin rash – raised red patches
- ▶ 3. Photosensitivity – unusually strong reaction to sun light, causing a rash or flare
- ▶ 4. Mouth or nose ulcers – usually painless
- ▶ 5. Nonerosive arthritis – inflammation in two or more joints.
- ▶ 6. Cardio-pulmonary involvement – inflammation of the heart lining and/or lungs
- ▶ 7. Neurologic disorder – seizures and/or psychosis
- ▶ 8. Kidney disorder – increased protein or clumps of red cells in urine
- ▶ 9. Blood disorder – anemia caused by damaged red cells, low white cells or low platelet count
- ▶ 10. Immunological disorder – when your immune system attacks healthy cells
- ▶ 11. Antinuclear antibodies (ANA) – positive blood test not induced by drugs

TREATMENT OF SYSTEMIC LUPUS

TREATMENT

1. There is NO cure for systemic lupus – BUT- the treatment of lupus has improved substantially over the past several years and survival rates are now close to that of the general population.
2. Treatment early in the course of the disease and appropriate follow up improves prognosis.

OF

3. There is NO one drug that treats ALL the manifestations of systemic lupus.
4. You generally treat what the lupus is doing, and target the organ affected.
5. There may be 10 people in a room with lupus and all 10 are on different medications for their disease.

LUPUS

6. But the goals of treatment are the same!
7. Goals of treatment:
 - Control symptoms like joint pain and fatigue
 - Reduce inflammation caused by lupus
 - Suppress your overactive immune system
 - Prevent flares and treat them when they occur
 - Minimize damage to organs

USE OF HYDROXYCHLOROQUINE IN SYSTEMIC LUPUS

ALL PATIENTS WITH
DOCUMENTED SYSTEMIC
LUPUS SHOULD BE ON
HYDROXYCHLOROQUINE
UNLESS THERE IS A
SPECIFIC REASON THY
CANNOT TOLERATE THE
DRUG.



- ▶ 1. Canadian STUDY: Hydroxychloroquine Study Group A randomized study of the effect of withdrawing hydroxychloroquine sulfate in systemic lupus erythematosus. *N Engl J Med* 1991; 324:150–154.
- ▶ 2. Tsakonas E, Joseph L, Esdaile J M, Choquette D, Senecal J L, Cividino A. *et al* A long-term study of hydroxychloroquine withdrawal on exacerbations in systemic lupus erythematosus. *Canadian Hydroxychloroquine Study Group. Lupus* 1998 ; 7:80–85.
- ▶ 3. Petri M. Hydroxychloroquine use in the Baltimore lupus cohort: effects on lipids, glucose and thrombosis. *Lupus* ; 15:16–22.
- ▶ 4. Fessler B J, Alarcon G S, McGwin G, Jr, Roseman J, Bastian H M, Friedman A W. *et al* Systemic lupus erythematosus in three ethnic groups. XVI: Association of hydroxychloroquine use with reduced risk of damage accrual, *Arthritis Rheum* 2005; 52:1473–1480.
- ▶ 5. Kasitanon N, Fine D M, Haas M, Magder L S, Petri M. Hydroxychloroquine use predicts complete renal remission within 12 months among patients treated with mycophenolate mofetil therapy for membranous lupus nephritis. *Lupus* 2006; 15:366–370
- ▶ 6. Effect of hydroxychloroquine on the survival of patients with systemic lupus erythematosus: data from LUMINA, a multiethnic US cohort (LUMINA I) Graciela S Alarcón, Gerald McGwin, Ana M Bertoli, Barri J Fessler, Jaime Calvo-Alén, Holly M Bastian, Luis M Vilá, and John D Reveille, for the LUMINA Study Group *Ann Rheum Dis.* 2007; Sep; 66(9): 1168–1172

TREATMENT OF SYSTEMIC LUPUS

1. Remember – you treat what the lupus is causing!
2. NSAIDs, Analgesics, Neuroleptics, Antidepressants, Physical therapy, etc.
3. Glucocorticoids – Corticosteroids – Steroids: Adjunctive and never the basis of long-term therapy.
4. Antimalarials – Hydroxychloroquine, Chloroquine
5. Biologic – B-lymphocyte Stimulating Inhibitor (Belimumab “Benlysta”)
B-Cell CD-20 Binding protein (Rituximab “Rituxan”).
6. Immunosuppressants/cytotoxic – methotrexate, azathioprine, mycophenolate, cyclophosphamide

TOXICITY OF TREATMENT

Group 2 – stomach ulcers, renal disease, liver disease, heart disease, etc.

Group 3 – wt. gain, increased glucose, HTN, rash, emotional changes, etc.

Group 4 – ocular toxicity, skin changes, decreased blood ct., cardiac, etc.

Group 5 – depression, infection risk, PML, allergic reaction, cancer, etc.

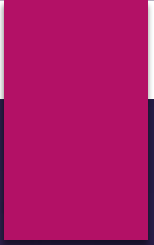
Group 6 - infection, decreased blood ct., liver and kidney, fertility, etc.

SYSTEMIC LUPUS

- Is quite treatable and your prognosis is good!!
- It is an autoimmune disease – so see a Rheumatologist early!
- Stay vigilant! Lupus is a constantly changing condition.
- Stay supportive!!

RHEUMATOLOGIST CREED

O*N My Honor*, I pledge, to the best of my ability, to strive to make an accurate diagnosis of any inflammatory or non-inflammatory autoimmune, rheumatologic condition, based on objective scientific data, in the earliest course of the disease as possible and to initiate safe and effective therapy, always seeking remission, limiting disease progression, preserving functional capacity and enhancing quality of life.



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QUESTIONS AND ANSWERS