

Lupus 101

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Lupus is a Systemic Autoimmune Disease

Systemic autoimmunity

SLE

RA

Sjogren's syndrome

Scleroderma

Inflammatory myositis

Vasculitis

Organ-specific autoimmunity

Autoimmune thyroiditis

Primary biliary cirrhosis

Autoimmune hepatitis

Type I diabetes

Multiple sclerosis

Myasthenia gravis

Systemic Lupus Erythematosus

- Inflammatory multisystem disease primarily seen in females (90%), but can be seen at any age in either sex
- Highly variable course and prognosis, ranges from mild to life threatening
- ~50% of patients can have severe internal organ involvement which can be life threatening without treatment
- Characterized by flares and remissions
- Associated with characteristic autoantibodies

Lupus history

- Lupus is the Latin word for wolf
- 1st used medically in the 10th century
- Described clinically in the 19th century
 - Butterfly rash in 1845
 - Arthritis in 1892
 - Nephritis in 1895 by Osler
- Serologic tests become available in the 20th century
 - LE cell in 1948
 - Lupus anticoagulant in 1952
 - ANA in 1954

From Dubois

What are the different forms of lupus?

Systemic Lupus Erythematosus

Discoid or Cutaneous Lupus

Drug-Induced Lupus

Neonatal Lupus

What lupus is not

AIDS

Cancer

Arthritis

Contagious

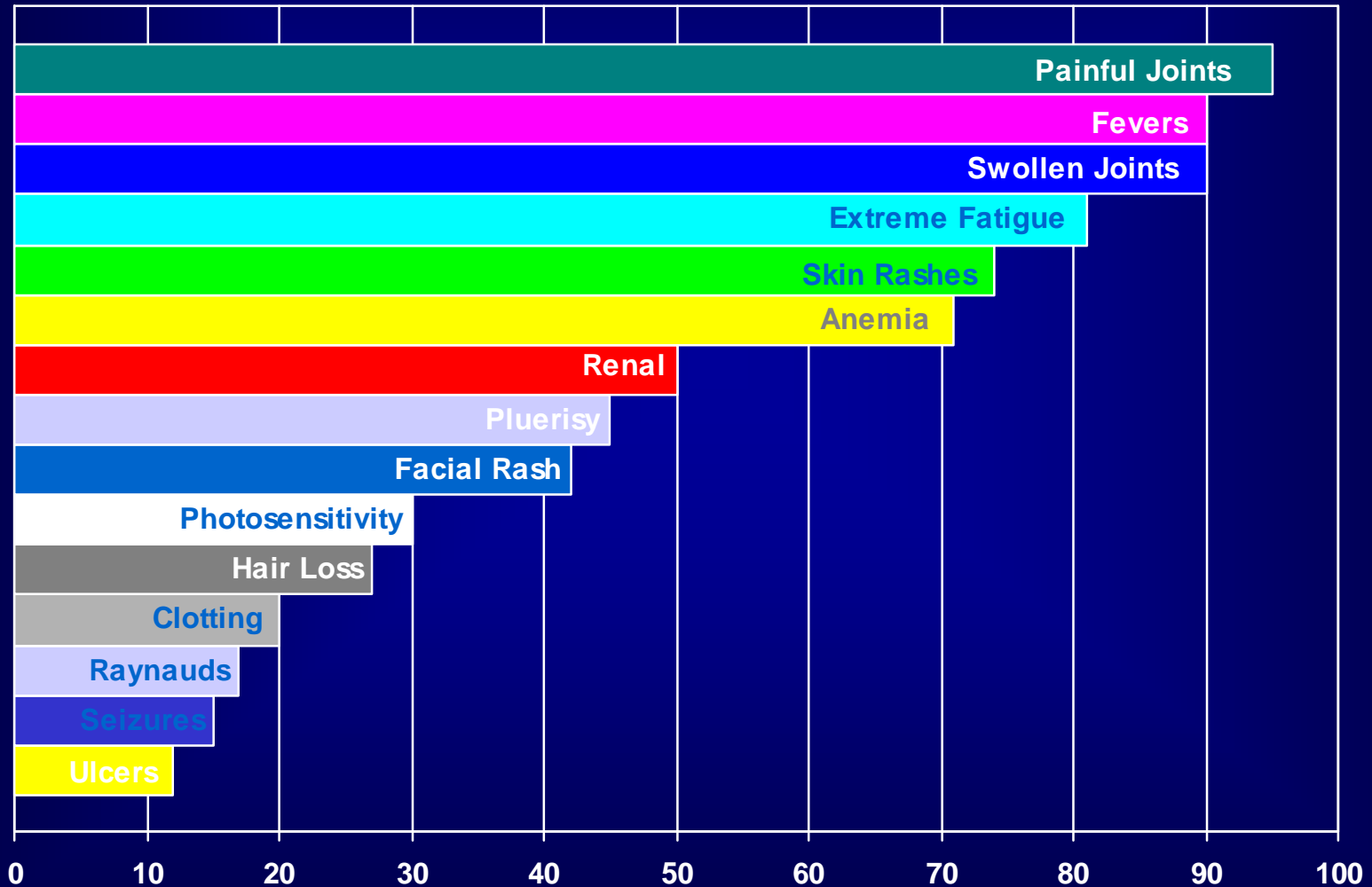
What are the symptoms of lupus?

- Painful swollen joints
- Unexplained fever
- Extreme fatigue
- Rashes
- Sensitivity to the sun
- Mouth Sores
- Hair loss
- Pale or purple fingers or toes from cold
- Swollen glands
- Headache and/or Depression
- Chest pain with deep breathing
- Low blood count

Other problems

- Repeated miscarriages
- Disease in organs
 - Kidney
 - Heart
 - Lungs
 - Brain and nerves

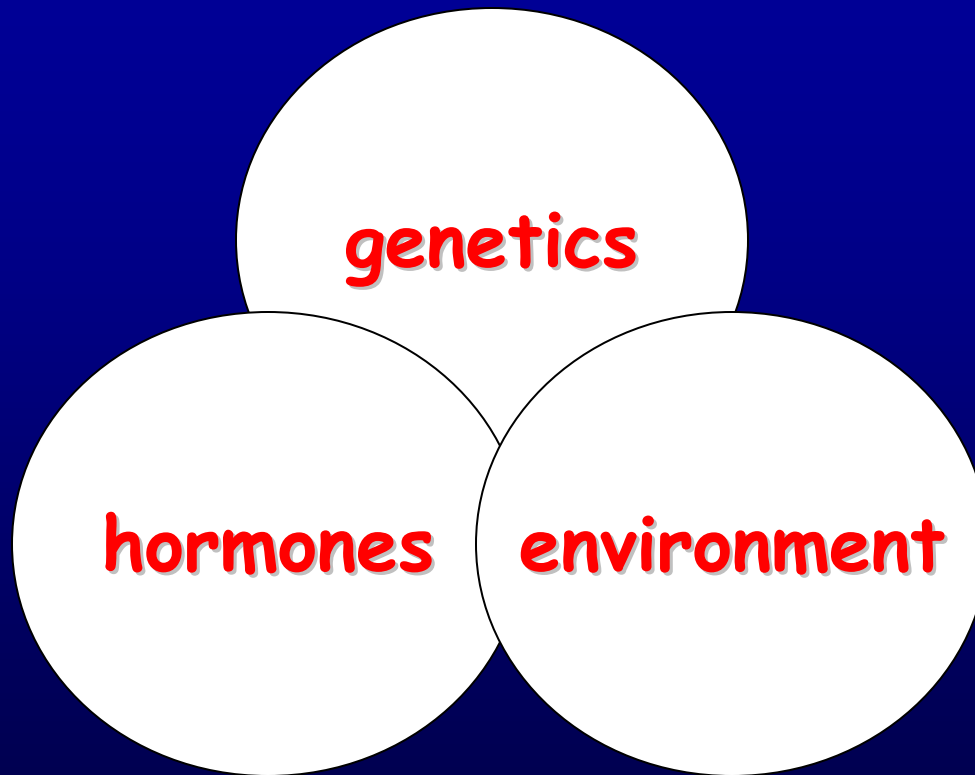
Lupus presenting symptoms



Who gets lupus?

- World-wide prevalence : 10-100/100,000
- Estimated frequency:
 - 1 per 700 white women
 - 1 per 245 black women
- The Lupus Foundation estimates that 1.5 million Americans have some form of the disease
- African-Americans > Caucasians (3x)
- Asian-American and Hispanics > Caucasians
- Age at diagnosis:
 - 16-55 years of age: 65% of cases
 - < 16: 20%
 - > 65: 15%
- 9/10 lupus patients are women

Cause



Triggers

Ultraviolet light

Stress

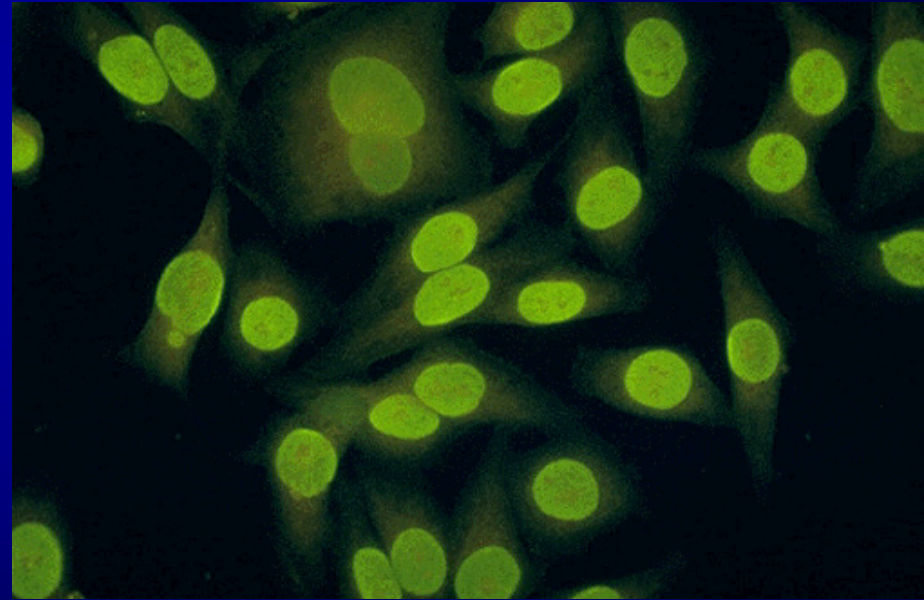
Medications

Infections

Hormonal Changes

SLE Diagnosis: The ANA

- ANA
 - Seen in 99% of SLE
 - Not specific for SLE
 - Seen in many inflammatory, infectious, and neoplastic diseases
 - Seen in 5% to 15% of normal persons



Where else can +ANA be seen?

- Normal individuals
- Individuals with family members who have SLE
- Other autoimmune diseases: rheumatoid arthritis, Sjogren's, scleroderma
- Infectious diseases: mononucleosis, endocarditis
- Organ specific autoimmune diseases: thyroid, liver
- Patients who will develop lupus in the future

Specific autoantibodies in SLE

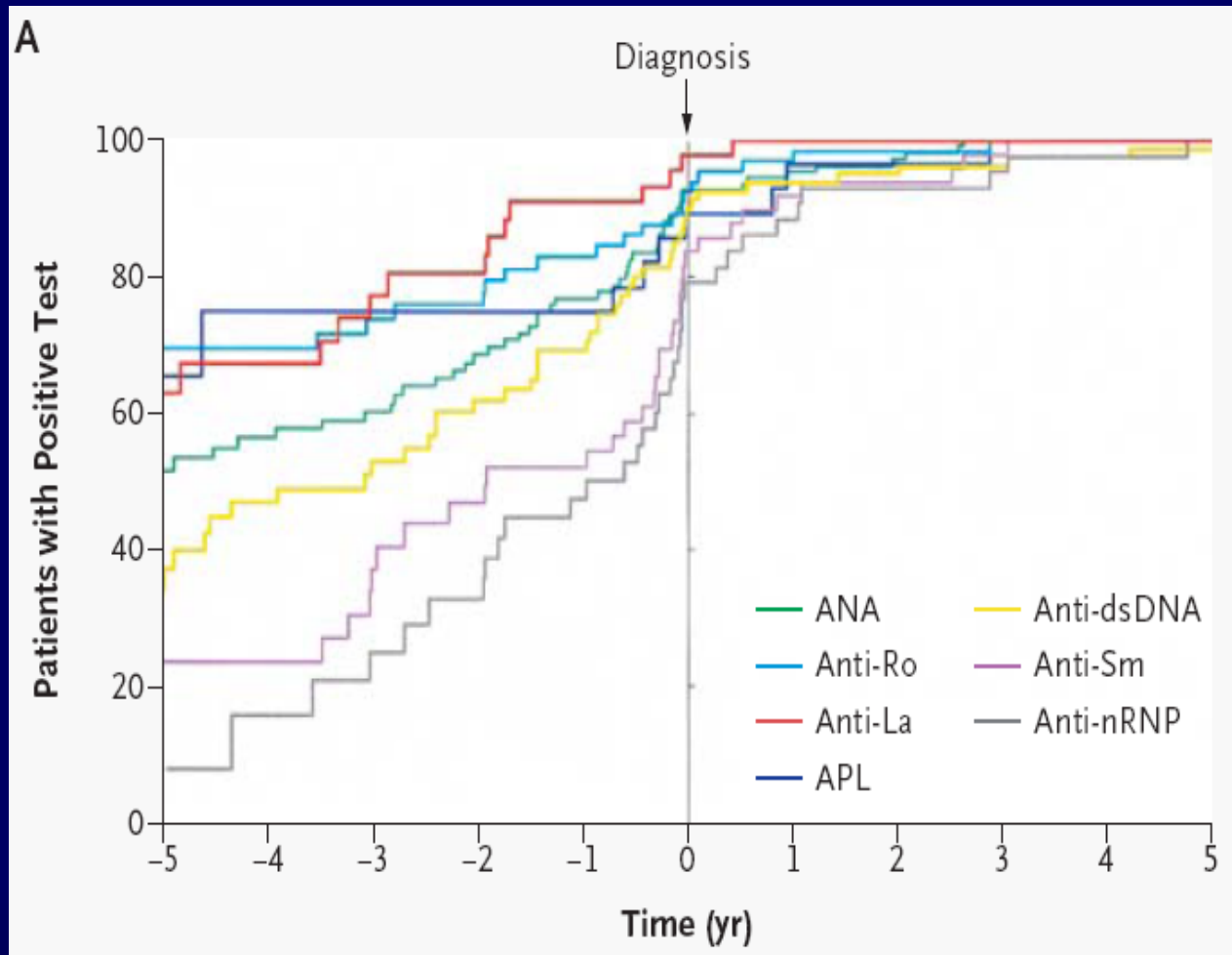
- **Anti-ds DNA**

- Seen in 40-60% of patients with SLE: the absence of anti-dsDNA does not exclude lupus!!
- Highly specific for SLE
- Low titer rarely seen in other inflammatory conditions
- Strongest clinical association is with nephritis

- **Anti-Sm (Smith)**

- Seen in 10% to 30% of SLE patients
- Highly specific for SLE

Autoimmunity before the onset of SLE



How do we diagnose lupus?

- Medical History (family history)
- Complete Physical Examination
 - Laboratory Tests
 - Skin or Kidney Biopsy

How do we diagnose lupus?

Skin criteria

1. Malar rash
2. Discoid Rash
3. Photosensitivity
4. Oral Ulcers

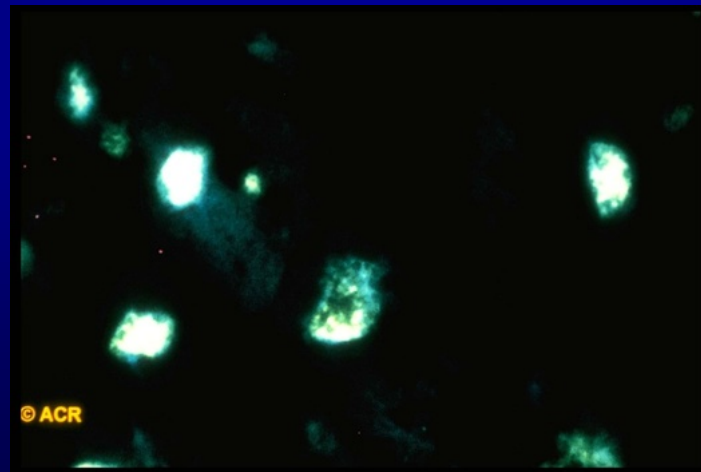
Systemic criteria

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

Lab criteria

9. Anti-nuclear antibody
10. Immunologic
11. Hematologic

*4 criteria simultaneously or serially for diagnosis



How is lupus treated?

Treating inflammation or autoimmunity

- Anti-inflammatory agents
- Antimalarials
- Immunosuppressive/cytotoxic agents

Other

- Prevention: management of cardiovascular risk, immunization, etc.
- Anti-thrombotic therapy
- Treating seizures
- Dialysis and kidney transplantation

The 'traditional treatment armamentarium'

FDA Approved drugs

- glucocorticoids
- hydroxychloroquine
- low dose ASA

'Off-label' but standard of care

- azathioprine
- cyclophosphamide
- NSAIDs

Immunosuppressives developed for other diseases

- mycophenolate mofetil methotrexate
- cyclosporin leflunomide
- tacrolimus fludarabine

Other aspects of treatment

* Sun Precautions

☾ Adequate Rest

🍴 Proper Nutrition

🏃 Moderate Exercise

🚭 Avoid Smoking

Treating Widespread Pain

Treating Depression and Anxiety

The benefits of hydroxychloroquine

- Decreased incidence and severity of flares of disease
- Low toxicity- q6 month eye exams but retinal toxicity is exceedingly rare

Not All Lupus Is the Same

Mild

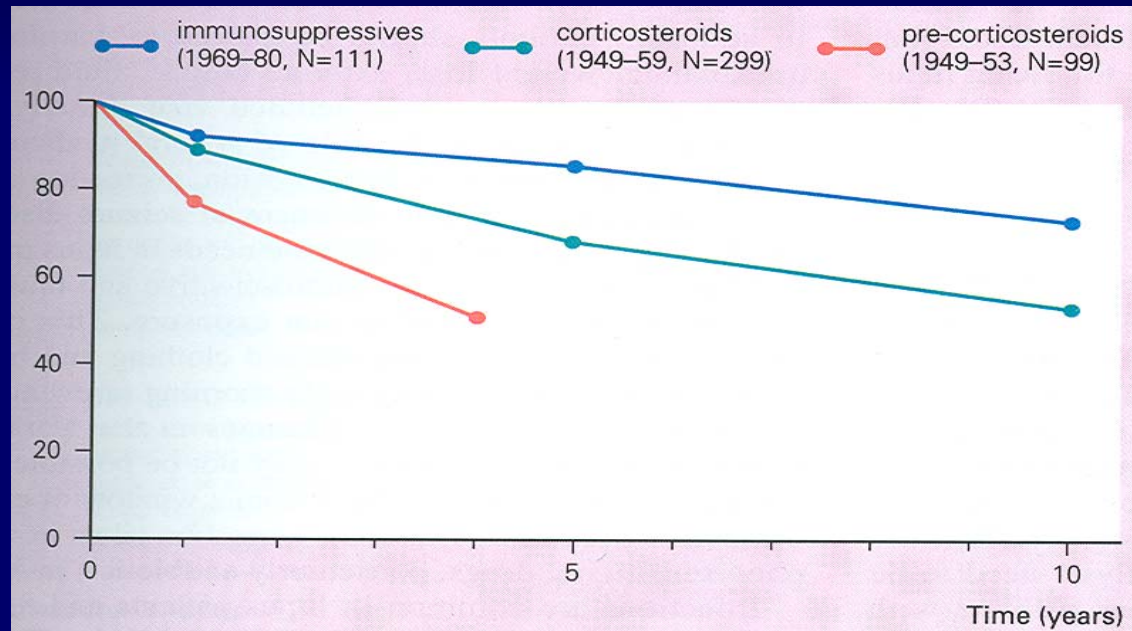
- Rash, oral ulcers, arthritis; confined to skin and joints
- Fever, fatigue

Severe

- Diffuse ulcerating rash
- Significant kidney disease
- Brain disease
- Very low platelet or red blood cell count
- Bleeding into the lungs

...and the picture can change

Prognosis



- Profound improvement in mortality...BUT
- 10-20 yr survival rates of 70-80% (3-5 fold worse than age matched controls)
- Death rates among black women with lupus are 3-fold higher than other populations
- 36% of deaths from lupus are in patients <45 years

Prognosis

With better methods of diagnosis and treatment, prognosis has improved in the last two decades

90% of people with lupus can expect to live a normal life span

Things to Remember Tomorrow

- SLE is a heterogeneous autoimmune disease
- SLE can be mild and is almost always treatable
- Although lupus can affect almost any part of the body, most people experience symptoms in only a few organs.
- Treatment must be directed at the whole person not just the disease.