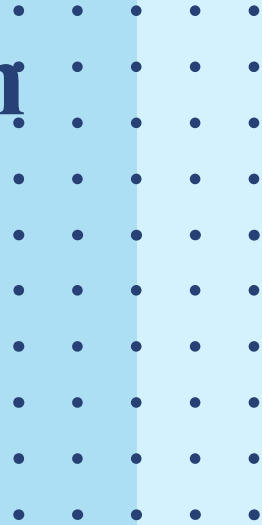
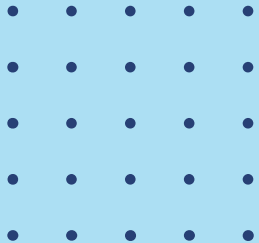


Kaleidoscope of Findings in Systemic Lupus Erythematosus

A Case Review





No disclosures

Case Review: History

A 27-year-old **female** with PMH significant for hypertension, anemia requiring transfusions, and type 2 diabetes mellitus presents to clinic with 4 days of **chest pain** and 1 day of **bilateral shin pain**.

HPI

- 15-lb weight loss
- ↓ appetite
- Epistaxis requiring multiple days of nasal packing

PMH

- Hypertension
- Anemia (hx transfusions)
- Type 2 diabetes mellitus
- Medications: lisinopril (40 mg/day), ferrous sulfate (325 mg/day), metformin (500 mg BID), Nexplanon

FH

- Diabetes mellitus
- Hypertension
- Congestive heart failure
- Myocardial infarction
- Menorrhagia (hx transfusions)

SH

- Denies use of alcohol, cigarettes, and illicit drugs
- From Hawaii
- Lives with brother and father

Case Review: Physical Exam

Vital Signs

BP: 157/68

HR: 128

RR: 18

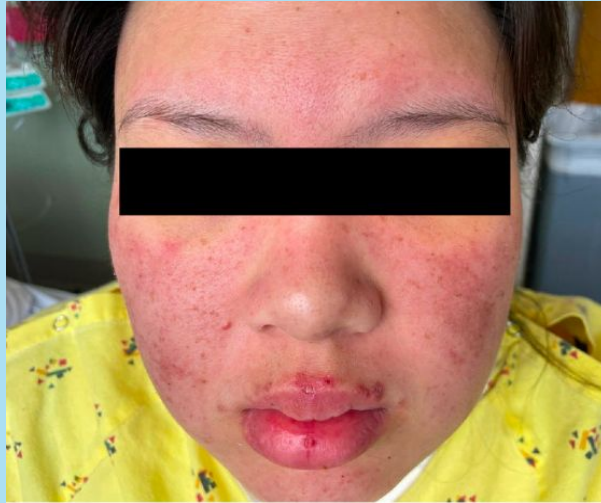
spO2: 98%

Temp: 101.2°F

Height: 5'2"

Weight: 185 lbs

BMI: 33.8



Thinning hair at crown of head

Erythematous patches on face/scalp



Aphthous ulcers

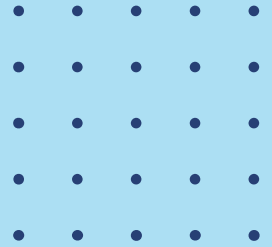
Petechiae on hard
palate



Bilateral LE
petechiae

Bilateral LE
edema

Case Review: Differential Diagnosis



SLE

Pulmonary Embolism

Immune Thrombocytopenic Purpura

Leukemia

Meningococcal Meningitis

Coxsackie Virus

Syphilis

Unspecified Vasculitis

Case Review: Labs & Imaging



WBCs: 2.9 ($4-10 \times 10^9/L$)
Platelets: 133,000 (150,000-350,000/uL)
Hemoglobin: 6 (F: 12-16 g/dL)
C3: 27 (55-120 mg/dL)
C4: 4 (15-45 mg/dL)



D-dimer: 4.6 ($< 0.5 \text{ ug/mL}$)
PTT: 37 (25-35 sec)
Creatinine: 1.36 (0.7-1.3 mg/dL)
ANA titer: 1:640, homogenous ($\leq 1:40$)
Anti-dsDNA: 1,637 ($< 10 \text{ IU/mL}$)

Urinalysis

- 2+ protein
- 3+ blood

Peripheral smear

- Leukopenia

Blood Culture

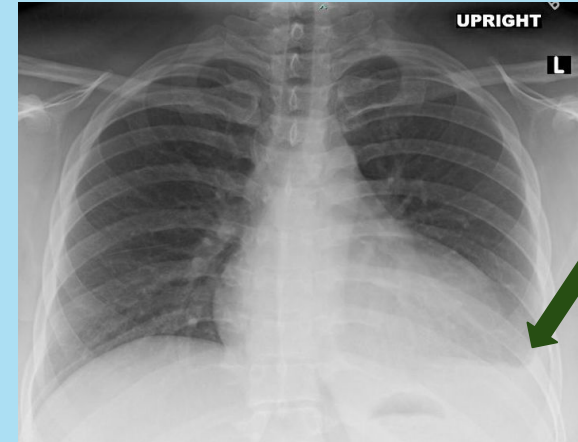
- NGTD x 2 days

CXR

- Decreased lung volumes
- Left pleural effusion

XR Tibia & Fibula

- Mild subcutaneous edema



Diminished
costophrenic
angle

What is Systemic Lupus Erythematosus?

Autoimmune disease

Heterogeneous
presentation

Any organ
system

Chronic or
cycles between
flares and
remission

Epidemiology

Incidence

5.1/100,000 people/year
(USA)

Prevalence

72.8/100,000 (USA)

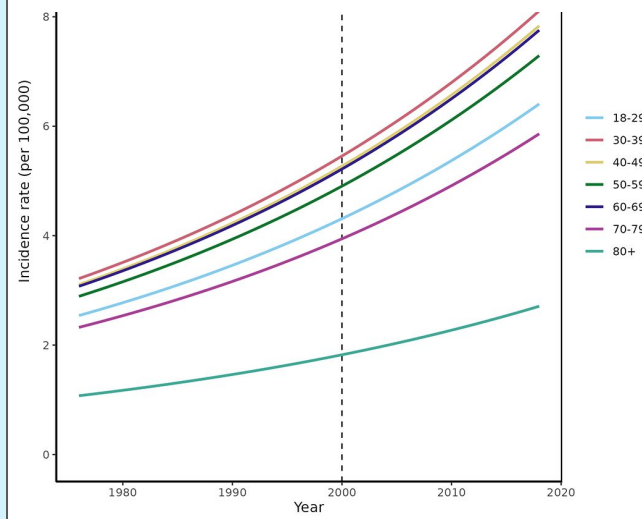
Sex

F > M (10:1)

Hawaii Demographics

Chinese > Hawaiian >
Filipino
(1975)

Trends in SLE incidence by age from
1976-2018 in Olmsted County, Minnesota



Duarte-García et al. (2022)

Vulnerable Populations

Women ages 15-44

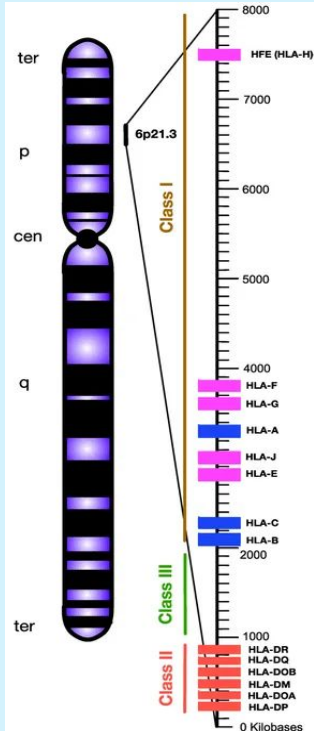
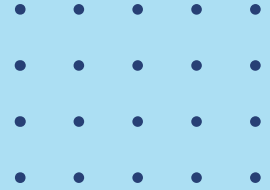
African American descent

Native American descent

Asian descent

Latin American descent

Etiology/Risk Factors

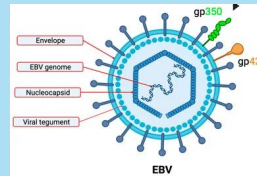


Genetic

- Concordance rates:
 - Monozygotic twins= 25-50%
 - Dizygotic twins= 5%
- HLA- DR 2, 3, 4, 8, 15
- Complement deficiency (C1q, C2, C4a)

Environmental

- UV exposure
- Tobacco
- Viral exposure (ex: EBV)

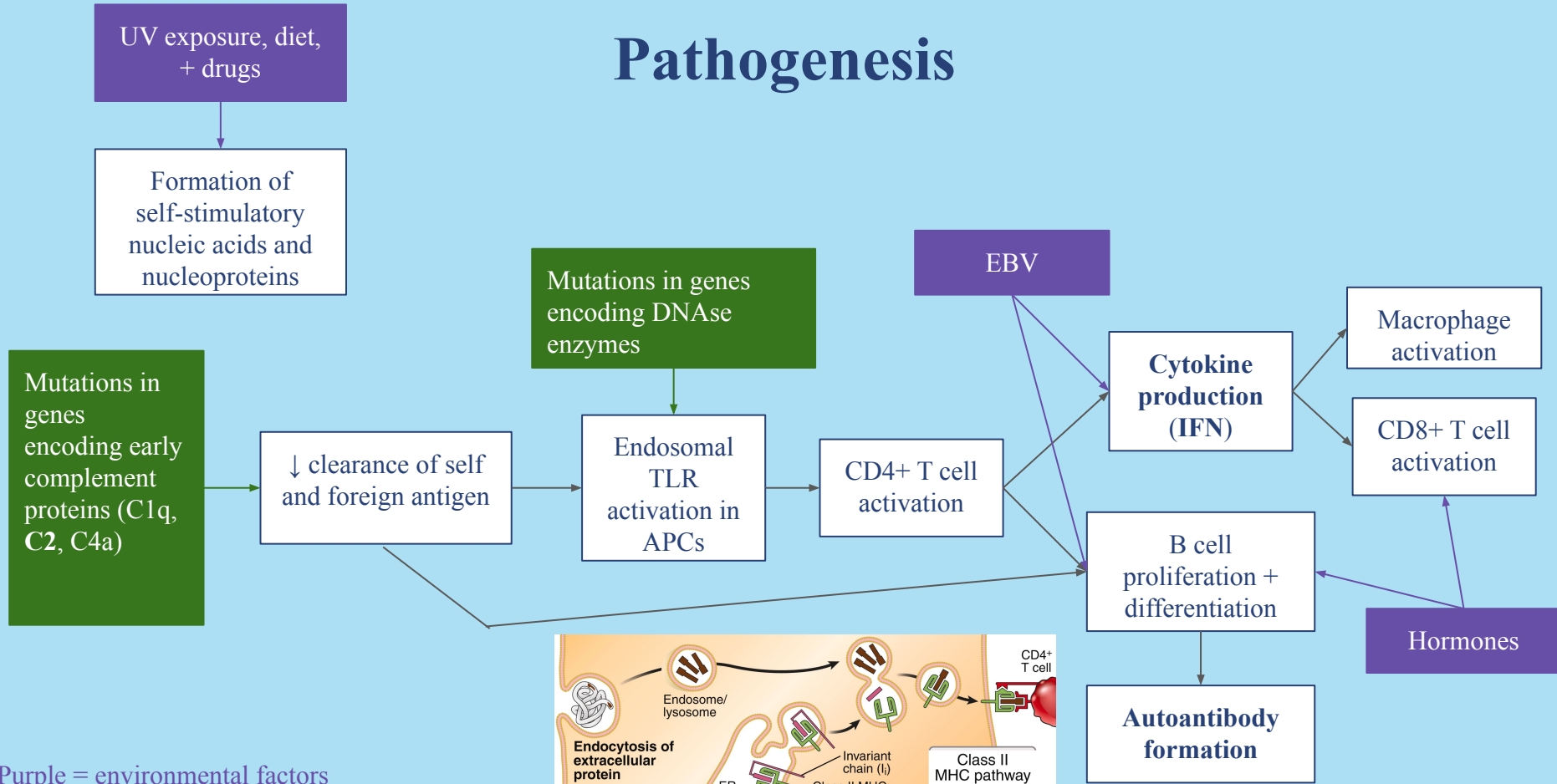


Iatrogenic

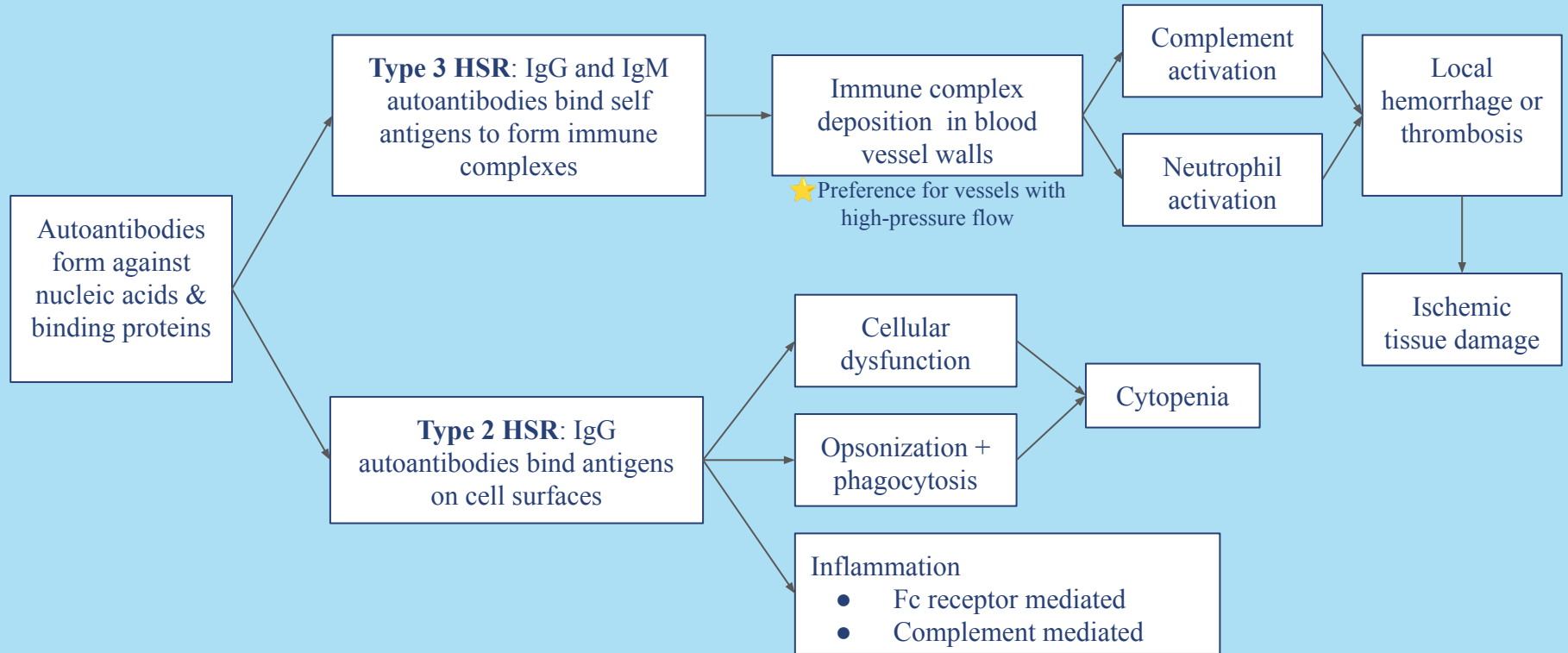
- Hormone therapy (OCPs, HRT)
- Procainamide
- Hydralazine
- Anti-TNF agents



Pathogenesis



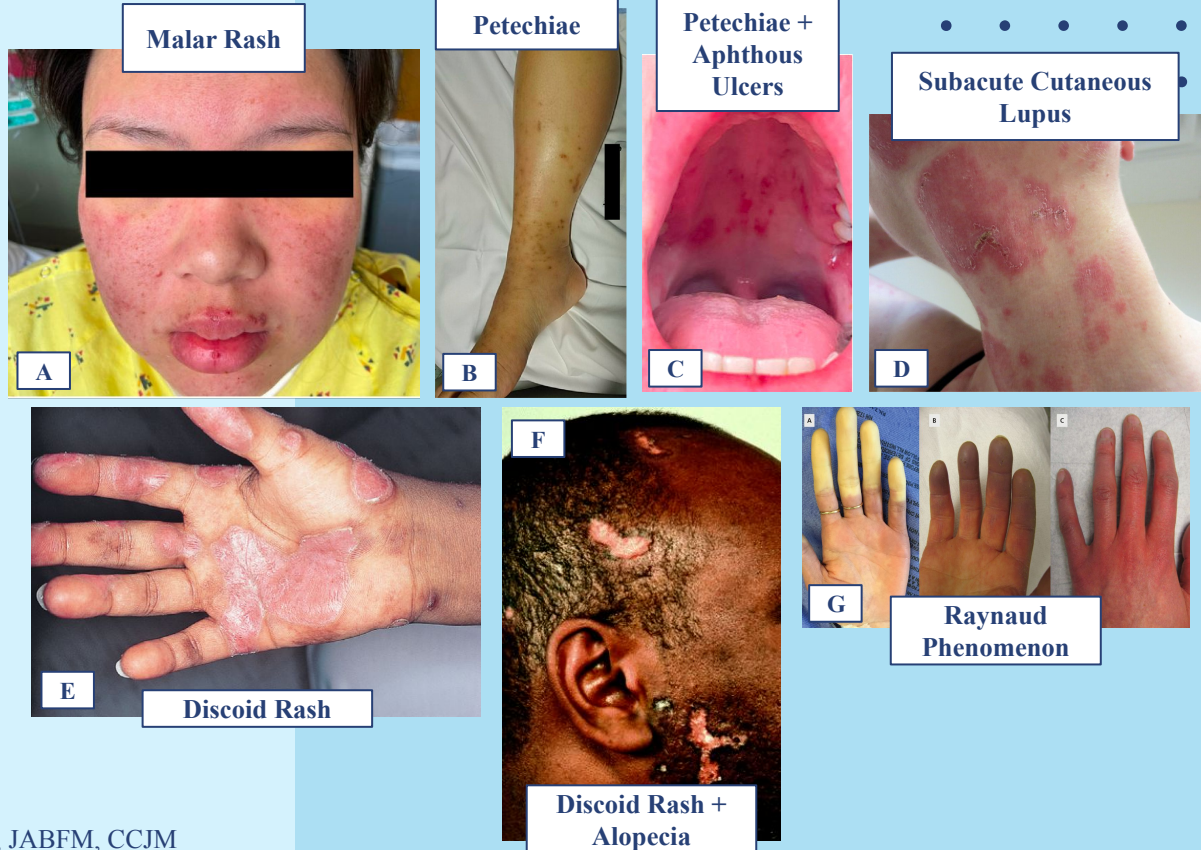
Pathophysiology



Symptoms

MANIFESTATION	APPROXIMATE FREQUENCY (%)
Cutaneous	88
Arthritis/arthralgias	76
Neuropsychiatric	66
Pleurisy/pericarditis	63
Anemia	57
Raynaud phenomenon	44
Vasculitis	43
Atherosclerosis	37
Nephritis	31
Thrombocytopenia	30
Sensorimotor neuropathy	28
Cardiac valvar disease	18
Pulmonary alveolar hemorrhage	12
Pancreatitis	10
Myositis	5
Myocarditis	5

Goldman L et al. (2024)

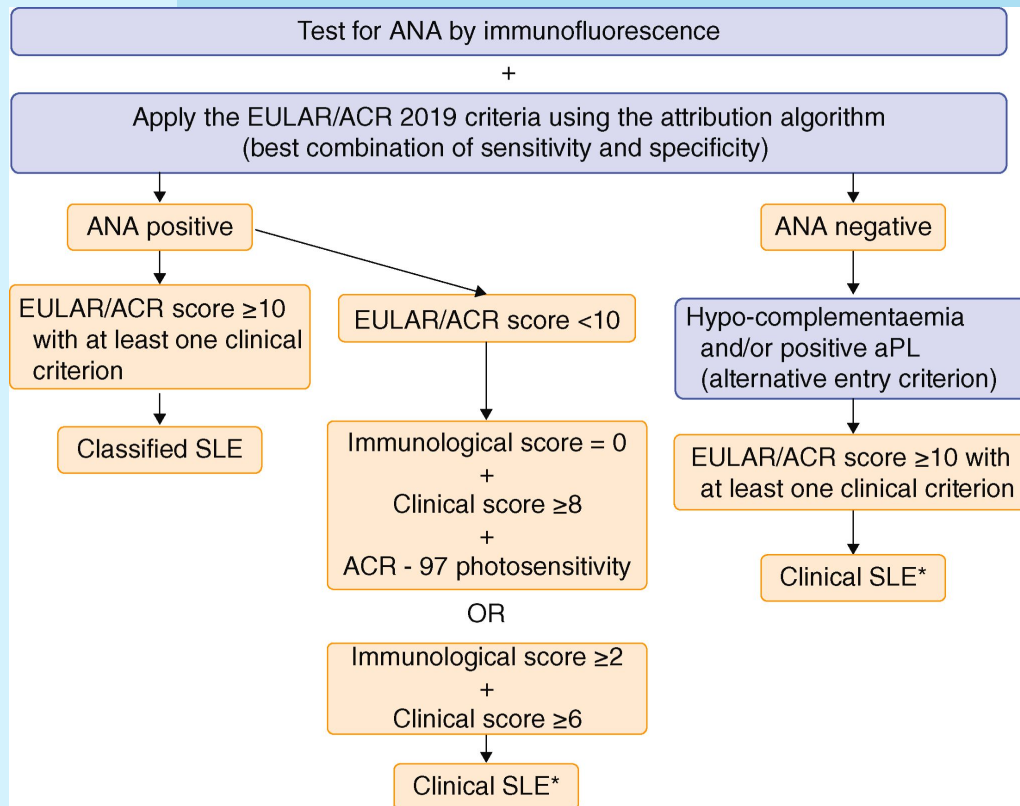


EULAR/ACR Criteria

Entry criterion			
Antinuclear antibodies (ANA) at a titer of $\geq 1:80$ on HEp-2 cells or an equivalent positive test (ever)			
↓			
If absent, do not classify as SLE If present, apply additive criteria			
↓			
Additive criteria			
Do not count a criterion if there is a more likely explanation than SLE. Occurrence of a criterion on at least one occasion is sufficient. SLE classification requires at least one clinical criterion and ≥ 10 points. Criteria need not occur simultaneously.			
Within each domain, only the highest weighted criterion is counted toward the total score ⁵ .			
Clinical domains and criteria	Weight	Immunology domains and criteria	Weight
Constitutional		Antiphospholipid antibodies	
Fever	2	Anti-cardiolipin antibodies OR	
Hematologic		Anti- β_2 GP1 antibodies OR	
Leukopenia	3	Lupus anticoagulant	2
Thrombocytopenia	4	Complement proteins	
Autoimmune hemolysis	4	Low C3 OR low C4	3
Neuropsychiatric		Low C3 AND low C4	4
Delirium	2	SLE-specific antibodies	
Psychosis	3	Anti-dsDNA antibody* OR	
Seizure	5	Anti-Smith antibody	6
Mucocutaneous			
Non-scarring alopecia	2		
Oral ulcers	2		
Subacute cutaneous OR discoid lupus	4		
Acute cutaneous lupus	6		
Serosal			
Pleural or pericardial effusion	5		
Acute pericarditis	6		
Musculoskeletal			
Joint involvement	6		
Renal			
Proteinuria $>0.5\text{g}/24\text{h}$	4		
Renal biopsy Class II or V lupus nephritis	8		
Renal biopsy Class III or IV lupus nephritis	10		
Total score:			
↓			
Classify as Systemic Lupus Erythematosus with a score of 10 or more if entry criterion fulfilled.			

Aringer et al. (2019)

Diagnosis



Goldman L et al. (2024)

Case Diagnosis

Entry criterion
Antinuclear antibodies (ANA) at a titer of $\geq 1:80$ on HEp-2 cells or an equivalent positive test (ever)
↓
If absent, do not classify as SLE If present, apply additive criteria
↓
Additive criteria
Do not count a criterion if there is a more likely explanation than SLE. Occurrence of a criterion on at least one occasion is sufficient. SLE classification requires at least one clinical criterion and ≥ 10 points. Criteria need not occur simultaneously.
Within each domain, only the highest weighted criterion is counted toward the total score.

Immunology domains and criteria	Weight
Antiphospholipid antibodies	
Anti-cardiolipin antibodies OR Anti- $\beta 2$ GP1 antibodies OR Lupus anticoagulant	2
Complement proteins	
Low C3 OR low C4	3
Low C3 AND low C4	4
SLE-specific antibodies	
Anti-dsDNA antibody* OR Anti-Smith antibody	6

TOTAL SCORE

27

CLASSIFIED AS
SLE

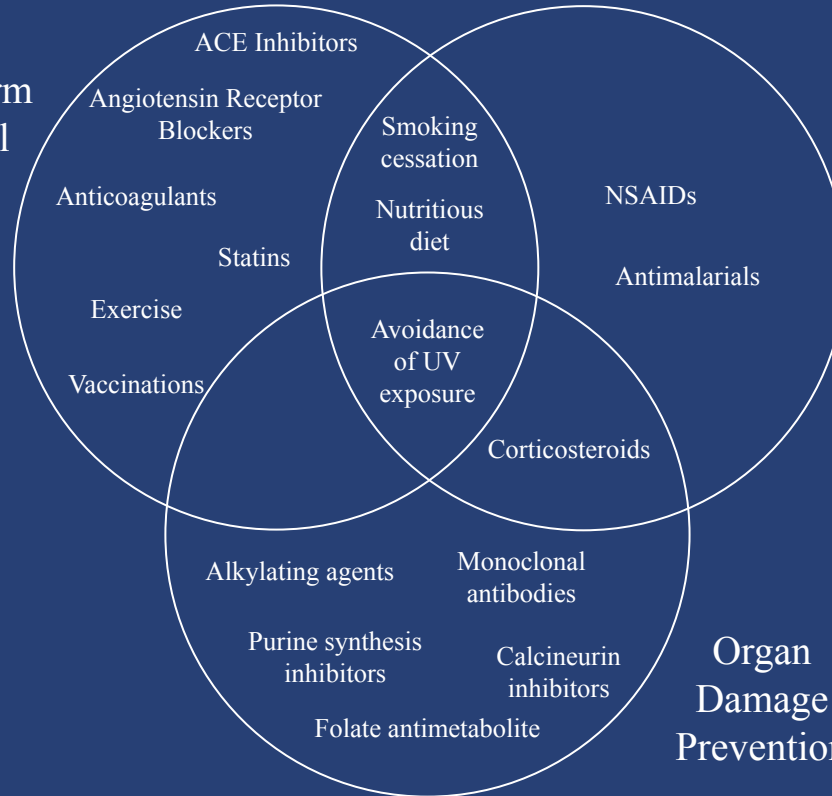
Clinical domains and criteria	Weight
Constitutional	
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Autoimmune hemolysis	4
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Delirium	2
Psychosis	3
Seizure	5
Mucocutaneous	
Non-scarring alopecia	2
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Subacute cutaneous OR discoid lupus	4
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Serosal	
Pleural or pericardial effusion	5
Acute pericarditis	6
Musculoskeletal	
Joint involvement	6
Renal	
Proteinuria $>0.5\text{g}/24\text{h}$	4
Renal biopsy Class II or V lupus nephritis	8
Renal biopsy Class III or IV lupus nephritis	10

Treatment

Long-Term
Survival

Reduction
of Disease
Activity &
Flare
Prevention

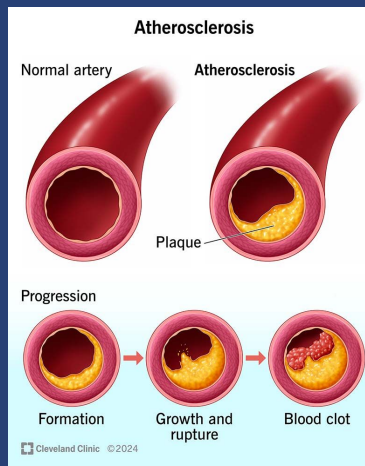
Organ
Damage
Prevention



Complications

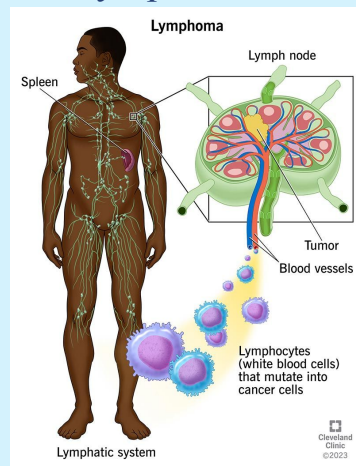
Atherosclerosis

- #1 cause of mortality in SLE



Malignancy

- **Hematologic:** non-Hodgkin lymphoma



Infection

- Linked to immune dysfunction + immunosuppressants

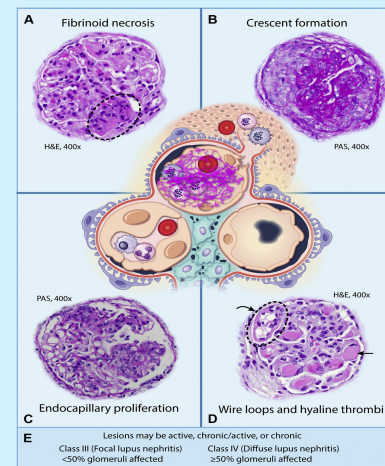
Table 1 Site of infection and microorganisms most frequently involved in systemic lupus erythematosus

Bacterial infections	
Respiratory tract	<i>Streptococcus pneumoniae</i> <i>Mycobacterium tuberculosis</i>
Urinary tract	<i>Escherichia coli</i> <i>Klebsiella</i> spp. <i>Pseudomonas</i> spp.
Skin and soft tissues	<i>Staphylococcus aureus</i>
Bacteremia/sepsis	<i>Escherichia coli</i> <i>Staphylococcus aureus</i> <i>Salmonella</i> spp.
Viral infections	
Skin	Herpes zoster
Respiratory tract	Cytomegalovirus
Gastrointestinal tract	
Central nervous system	
Lupus flare-like manifestations	
Cervix	Human papillomavirus
Fungal infections	
Upper gastrointestinal tract	<i>Candida</i> spp.
Genitourinary tract	
Respiratory tract	<i>Pneumocystis jirovecii</i>
Central nervous system	<i>Cryptococcus neoformans</i>
Respiratory tract	

Danza & Ruiz-Irastorza (2013)

Renal Disease

- Lupus nephritis



Wenderfer & Eldin (2019)

Prognosis

- <10% of patients achieve prolonged, complete remission
- Predictors of high disease activity
 - High anti-dsDNA antibodies
 - African American, Latin American, or Chinese ethnicity
 - No health insurance
 - Low social support

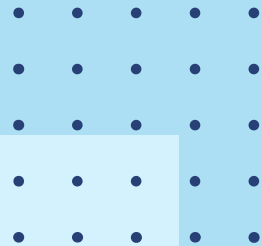
Causes of Death

Infection
Active disease complications

Cardiovascular disease
Treatment complications
Long-term organ damage

Early disease

Late disease



Thank you!

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