American College of Rheumatology

Empowering Rheumatology Professionals

RURAL HEALTH OUTREACH: PROVIDER EDUCATION ON LUPUS

Building connections between healthcare providers and improving the diagnosis and treatment of lupus

the lupus initiative

Eliminating Health Disparities in Lupus

Welcome.

We are excited to speak with you today about lupus.

Before we get started…

ACCREDITATION STATEMENT

The American College of Rheumatology is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to provide continuing medical education for physicians.

USE OF PROFESSIONAL JUDGMENT

This activity, including all educational links, is intended to be used as a tool to assess the base knowledge of the learner. The information presented relates to basic principles of diagnosis and therapy, and is meant in no way to substitute for an individual patient assessment based upon the healthcare provider's examination of the patient and consideration of laboratory data and other factors unique to the patient.

DRUGS AND DOSES

When prescribing medications, the physician is advised to check the product information sheet accompanying each drug to verify conditions of use and to identify any changes in drug dosage schedule of contraindications.

ACR DISCLOSURE STATEMENT

The American College of Rheumatology is an independent, professional organization that does not endorse specific procedures or products of any pharmaceutical/biotech concern.

SUPPORT

This project is supported by the Centers for Disease Control and Prevention of the U.S. Department of Health and Human Services (HHS) as part of a financial assistance award totaling $7.5 million over 5 years with 100 percent funded by CDC/HHS. The contents are those of the author(s) and do not necessarily represent the official views of, nor an endorsement, by CDC/HHS, or the U.S. Government.

PRESENTER REPORTED DISCLOSURES

[To be filled in by AHEC or presenter]

Consent and pre/post assessment

Participation in the assessments is voluntary and used to evaluate the effectiveness of the session.

No individual data will be shared as a part of this project. We will use your unique identifier only to match the assessments for analysis.

Final data will be reported in aggregate form.

1

CONSENT
(BEFORE SESSION)

- Sign and complete consent form if you would like to participate in the evaluation
- We **must** have your consent to use your evaluation data

2

PRE-ASSESSMENT
(BEFORE PRESENTATION)

- Multiple choice and true/false knowledge questions
- Self-efficacy questions
- Demographic questions

3

POST-ASSESSMENT
(AFTER PRESENTATION)

- Repeat of pre-assessment knowledge questions
- Additional qualitative questions regarding usefulness of session

Goals of Small Group Provider Sessions on Lupus

1. Build sustained connections between healthcare providers and a rheumatologist in areas and communities in which patients are underserved by rheumatologists.

2. Raise awareness among health care providers of lupus signs and symptoms, what to do if lupus is suspected and when to elevate lupus in the differential diagnosis when presented with individuals in groups at high risk for lupus.

Learning objectives

After this presentation, you should:

Recognize the signs and symptoms of lupus.

Know when to effectively refer a suspected lupus case to a rheumatologist.

Know how to initiate a work-up for lupus.

Have increased knowledge about lupus epidemiology, health disparities and disease characteristics.

A lupus diagnosis can take as long as two or more years and include visits to three or more health care providers.

Why is the diagnosis of lupus so challenging?

LUPUS IS:

- the great masquerader
- can mimic other conditions like viral syndromes, malignancies, allergic reactions and stress
- sometimes associated with depression or fibromyalgia

SYMPTOMS MAY BE VAGUE, INCLUDING:

- fatigue
- achiness
- stiffness
- low-grade fever
- swollen lymph nodes
- rashes

SYMPTOMS MAY:

- develop slowly
- come on suddenly

A lupus diagnosis can take as long as two or more years and include visits to three or more health care providers.

This delay in getting diagnosed can be devastating for a person with lupus.

THIS COULD LEAD TO:

- Organ failure
- a five-fold increased risk of death

With an early diagnosis, the chances of a person with lupus living a full life with a manageable, chronic disease are increased.[3]

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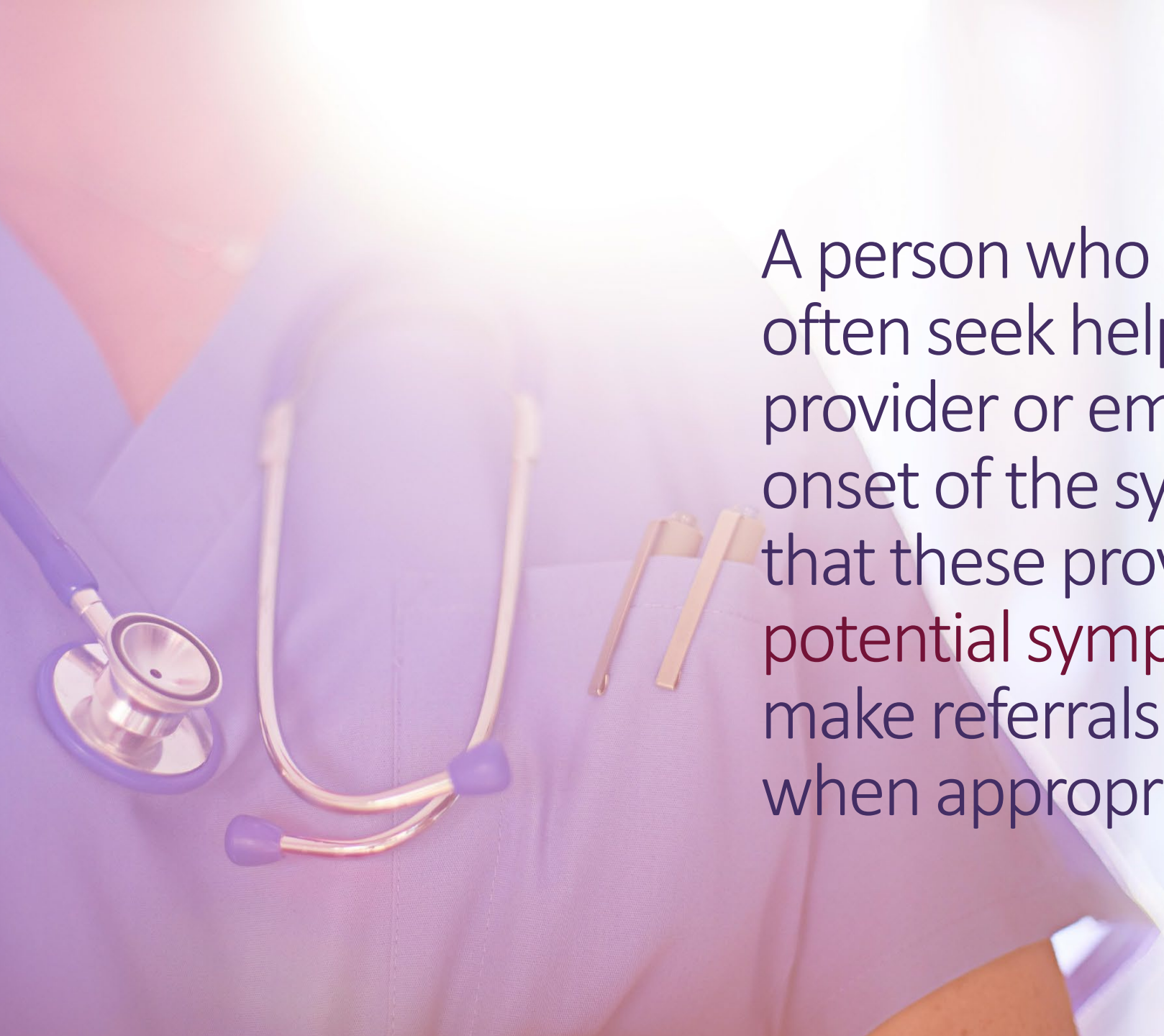


A person who is feeling sick will often seek help from a primary care provider or emergency room at the onset of the symptoms. It is crucial that these providers recognize potential symptoms of lupus and make referrals to rheumatologists when appropriate.

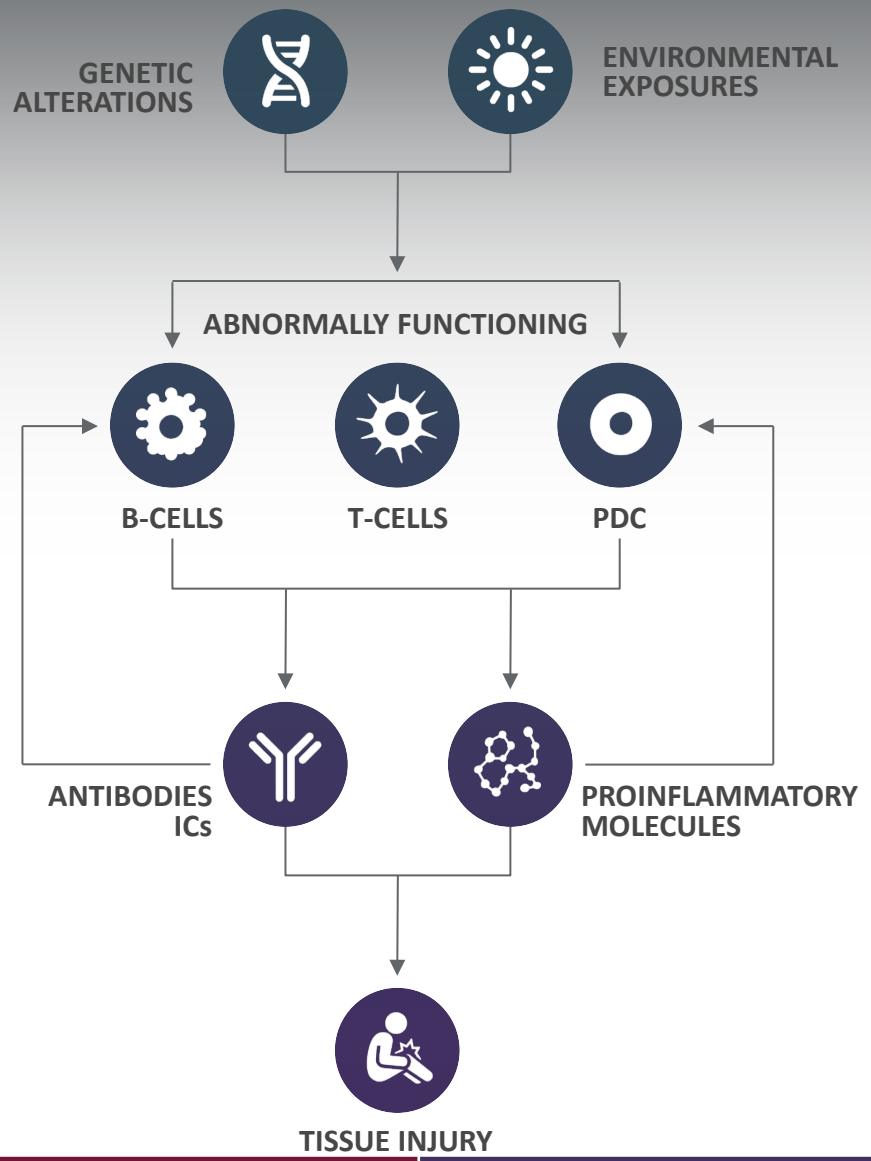
It is our mission to educate primary care providers about lupus, so people with lupus can get the correct referral, diagnosis and treatment they need.

About systemic lupus erythematosus (SLE)

- Lupus is an inflammatory, multi-system, autoimmune disease of unknown etiology with clinical manifestations that can change frequently and unexpectedly and suggestive laboratory manifestations.
- Lupus can be mild, severe and anything in between.
- The diversity of clinical symptoms is broad, and all organ systems are vulnerable.
- Lupus is characterized by periods of flare and remission and can culminate in irreversible, end-organ damage.

Pathogenesis of lupus

- Autoimmunity is an altered immune homeostasis that leads to autoreactivity, immunodeficiency and malignancy.
- Immune dysregulation leading to autoreactivity and autoantibodies in lupus occurs in different phases and likely represents the untoward effects of environmental triggers on the genetically susceptible host.



What are the demographics of lupus and populations at highest risk?

Epidemiology of lupus

Prevalence 2–140/100,000 worldwide but as high as 207/100,000
Incidence 1–10/100,000

Women are **nine times** more likely to develop lupus than men.[6]

African American and Hispanic/Latino women are more likely to develop lupus at a **younger age** and to have more severe symptoms at onset.[5]

Black, Latino, Asian and American Indian/Alaskan Native women have the **highest prevalence.**[6]

People with **lower incomes** are less likely to receive recommended care, and poverty is associated with poor outcomes.[5]

African American and Hispanic/Latino women with lupus have mortality rates at least **three times** as high as white women with lupus.[5]

African American women have a **three to six times** higher risk than white women.[2]

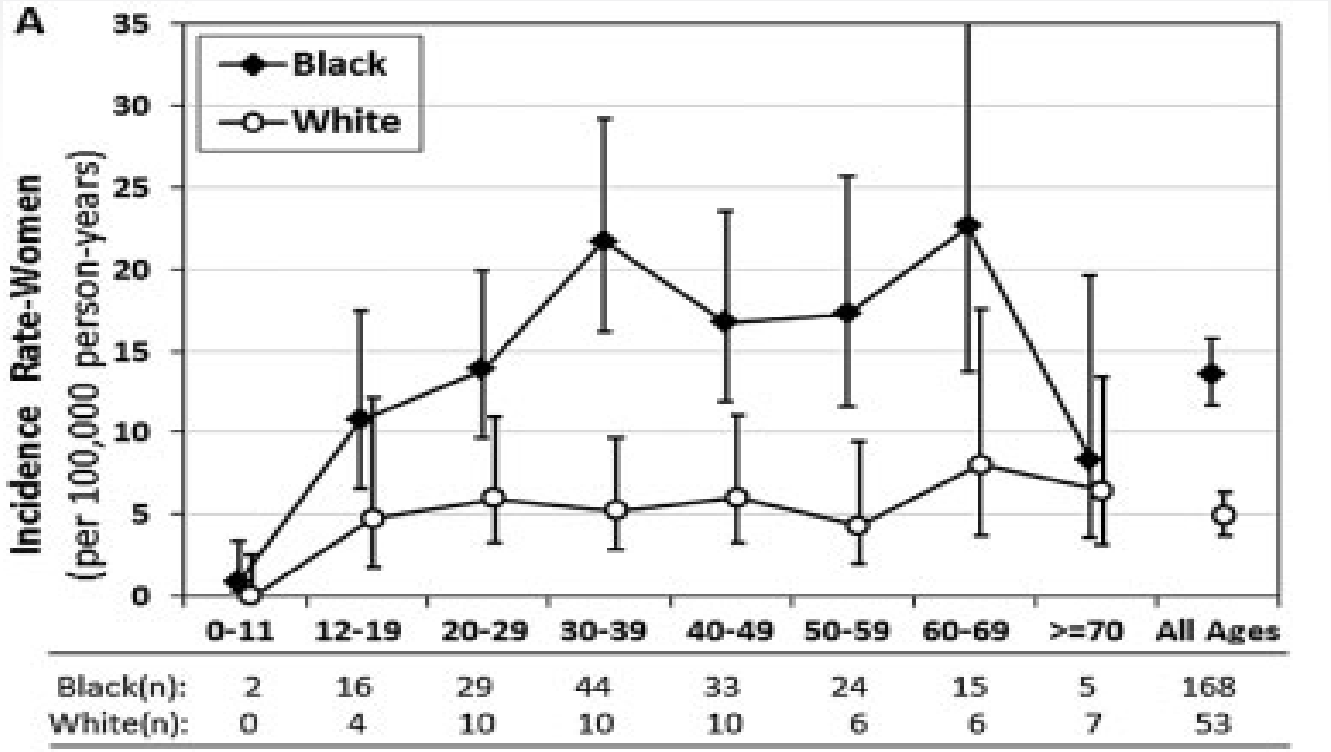
Affects mainly women in their **reproductive years.**[6]

The risk for Latino and Asian Women is between **1.4 and 1.7 higher** than for white women.[2]

Lupus affects up to **1 in 250** African American women in the United States.[6]

Disparities in Lupus Disease Burden

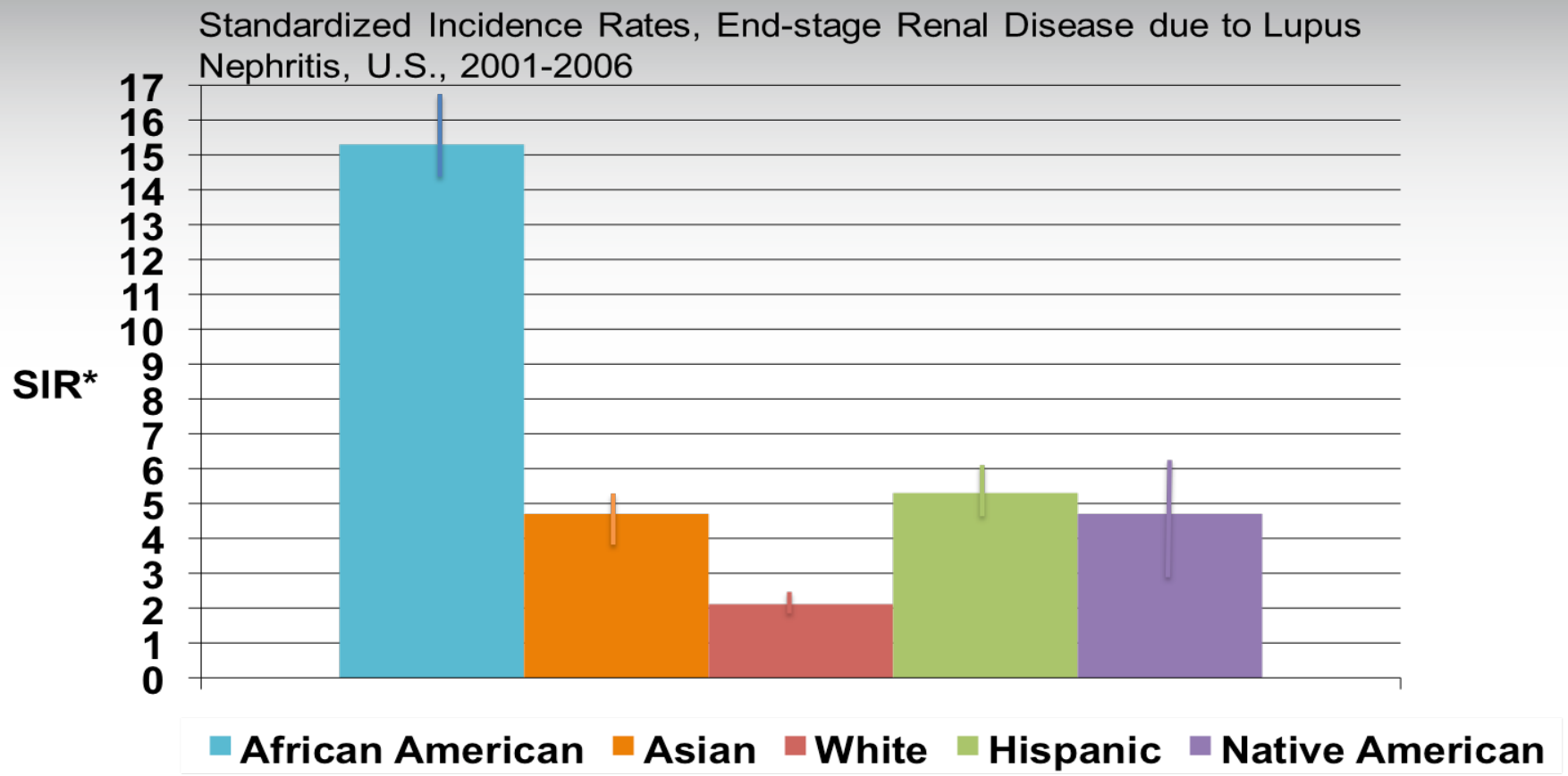
Racial/ethnic minorities are more likely to develop lupus at a younger age and to have more severe symptoms at onset.[7]



Other health disparities in lupus include[4]:

- Low income individuals less likely to receive recommended care
- Poverty associated with poor outcomes

Disparities in Lupus Outcomes: Renal[8]

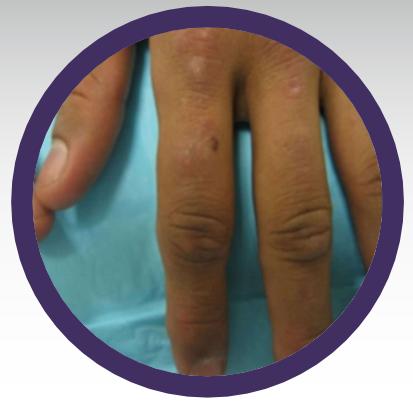


* Standardized Incidence Rate: end-stage renal disease cases/million person-years

Let’s take a look at some of the symptoms of lupus.

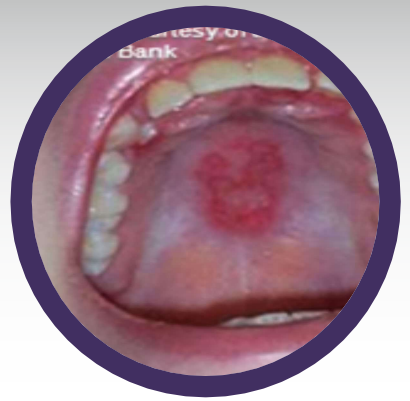
Here are some visible/external manifestations of lupus.

(When looking for skin conditions, keep in mind that skin conditions look different on diverse skin tones.)

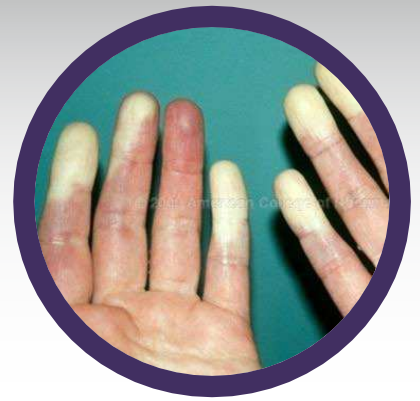


Synovitis

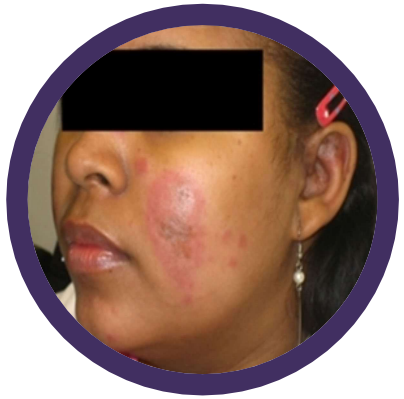
Malar rash



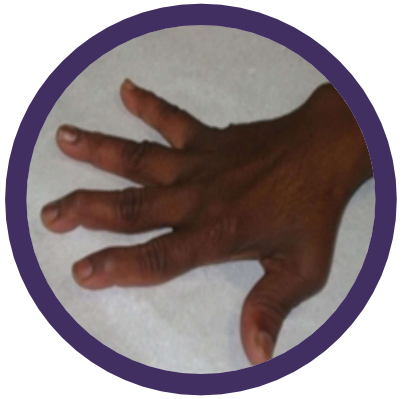
Painless oral ulcer



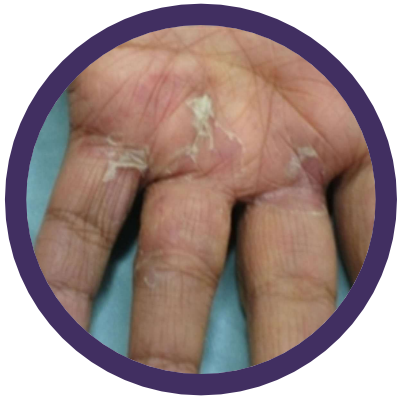
Raynaud's Phenomenon



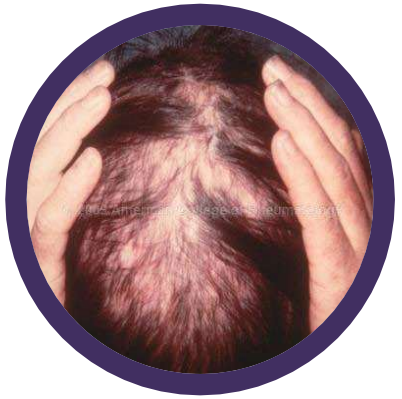
Discoid rash



Jaccoud's arthropathy



Vasculitis



Alopecia

Here are some lupus symptoms that make diagnosis difficult, because they can be indicators of so many conditions:

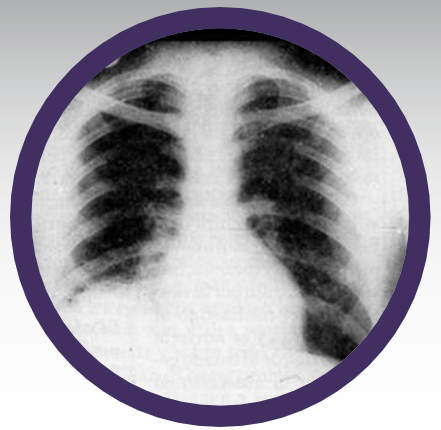
Achiness, headache

Fatigue

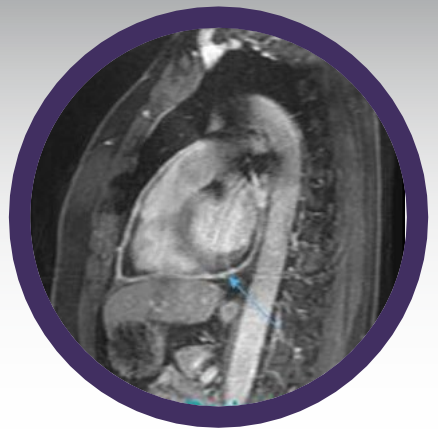
Memory thief / brain fog

Depression

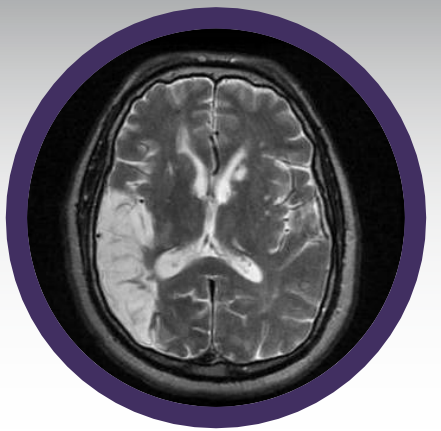
Here are some internal manifestations of lupus.



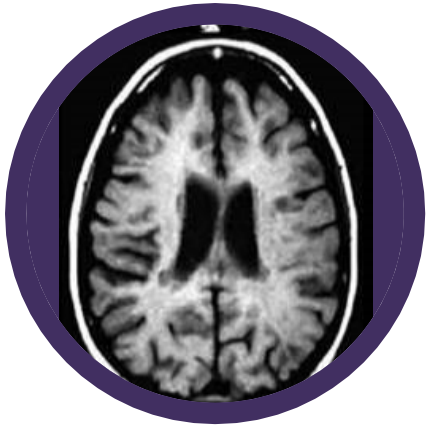
Serositis



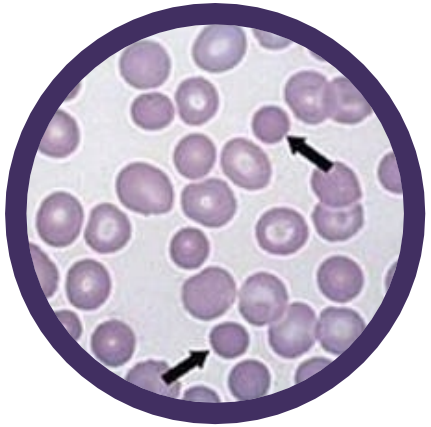
Pericardial effusion



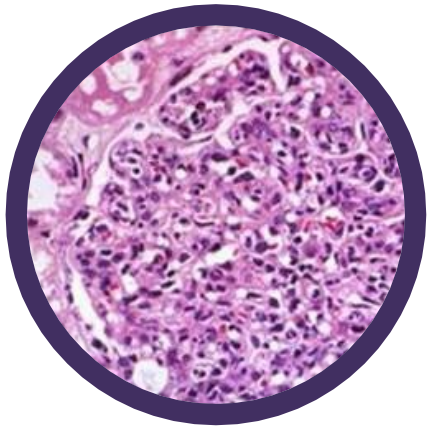
Cerebral infarct



Brain atrophy

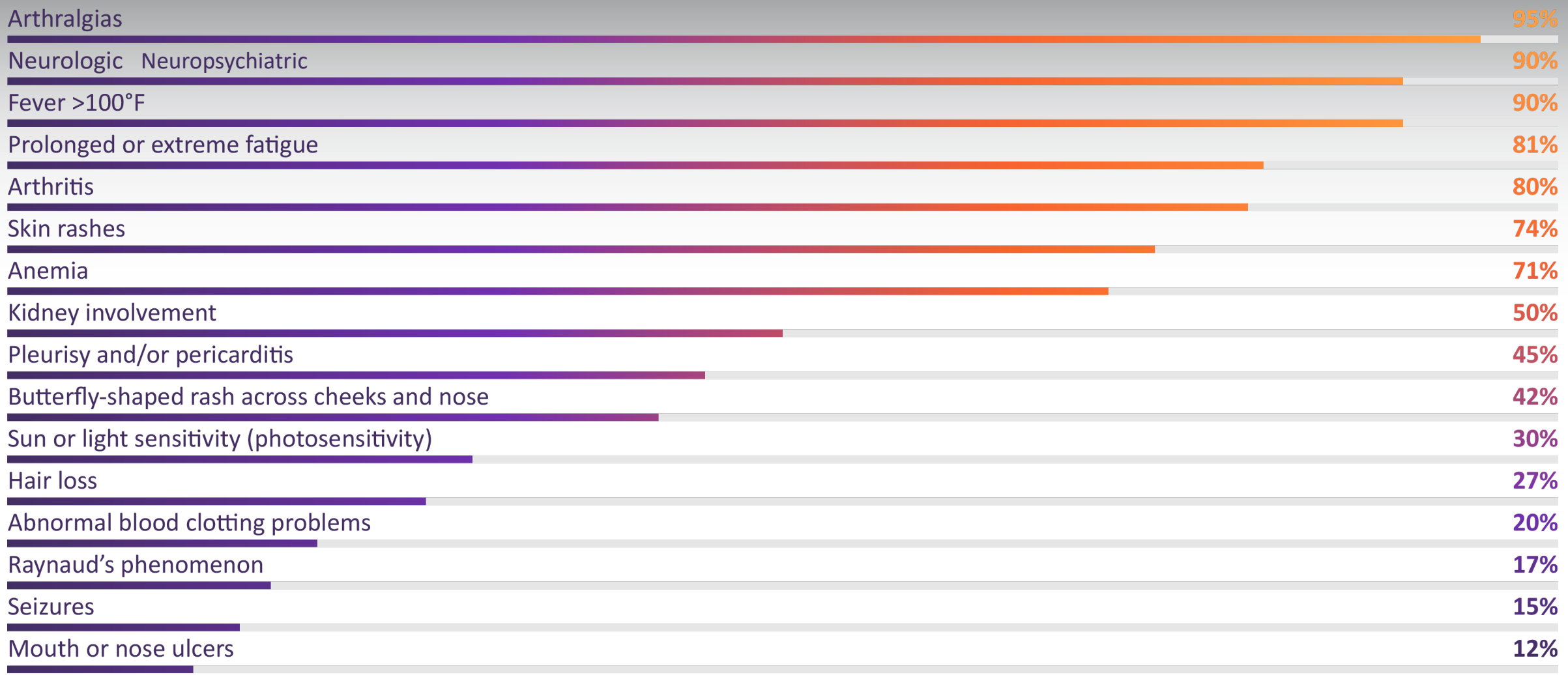


Spherocytes



Glomerulonephritis

Signs and symptoms



Unfortunately, there is no gold-standard diagnostic test for lupus.

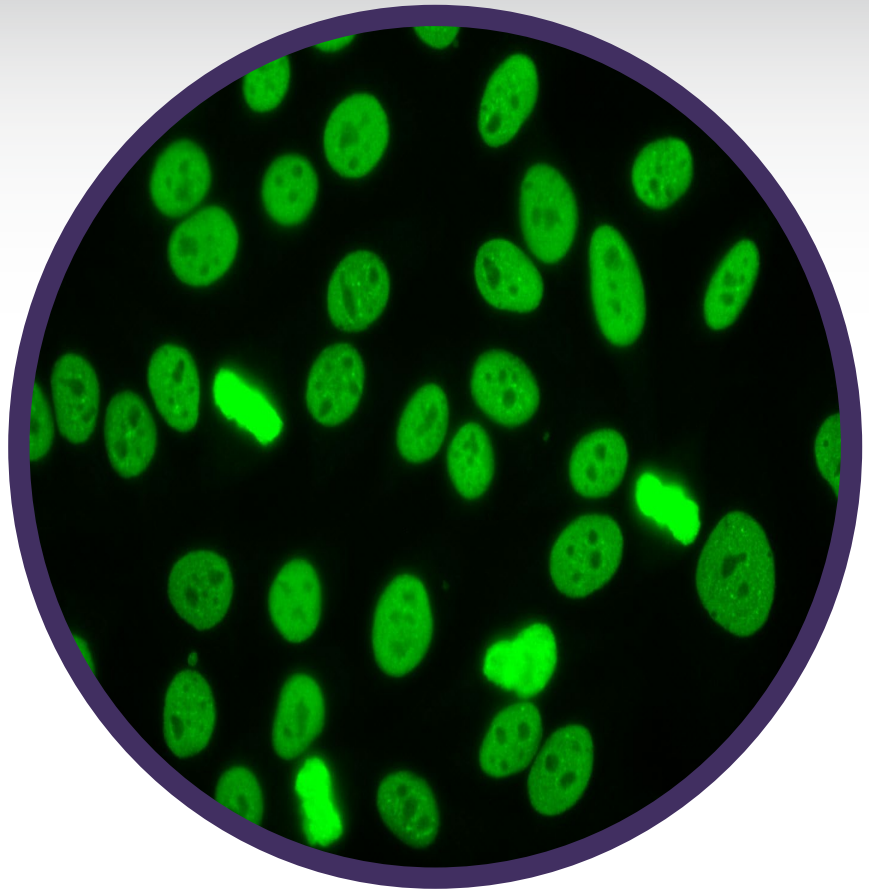
Clues to suspect SLE

- Presentation is often with arthritis and fatigue
- DDx might include RA, other connective tissue disease, other skin diseases, fibromyalgia, and others
- Common SLE clues:
  - Rashes, alopecia, oral ulcers, serositis
  - Leukopenia, lymphopenia
  - Proteinuria, RBCs/casts in urine

The most common screen is for ANA—antinuclear antibodies.

The vast majority of women with lupus test positive for ANA, but a positive ANA test does not mean the person has lupus. A positive ANA is also seen in the general population without known disease.

ANA could also indicate scleroderma, Hashimoto's thyroiditis, idiopathic pulmonary fibrosis and other chronic conditions.



What do most lupus patients have in common? antinuclear antibodies (ANA).

- Autoantibodies against various components of the cell nucleus.
- Present in many autoimmune disorders as well as some healthy subjects.
- Sensitive (not specific for SLE)
- Because of low specificity, ANA usefulness increases if the pretest probability for lupus is high; i.e., the patient has symptoms and signs that can be attributed to SLE.
- Because of the high sensitivity of the ANA, a patient with negative ANA is unlikely to have lupus even when her/his clinical presentation is suggestive of lupus.

If you suspect lupus, you can order these tests:

CBC WITH DIFFERENTIAL, UA, RENAL FUNCTION PANEL

- Urine protein/creatinine ratio if any proteinuria.

ANTINUCLEAR ANTIBODY (ANA)

- Positive in vast majority of patients with SLE.
- Beware false positives!
- Higher titer more likely to be clinically significant.
- If ANA is positive, consider additional autoantibodies.
- Anti-dsDNA and/or anti-Sm – more specific but less sensitive.

COMPLEMENTS: C3, C4

- Often but not always low in active disease.
- Acute phase reactants: elevated in other causes of inflammation (infection).

Autoantibodies in lupus

ANTIBODIES	LUPUS SPECIFICITY	LUPUS SENSITIVITY
ANA	● ○ ○ Low	● ● ● High
Anti-dsDNA	● ● ● High	● ● ○ Intermediate
Anti-Sm	● ● ● High	● ○ ○ Low
Anti-RNP	● ○ ○ Low	
Anti-SSA	● ○ ○ Low	
Anti-SSB	● ○ ○ Low	
Antiphospholipid	● ● ○ Intermediate	● ● ○ Intermediate[5]

So how do you determine whether to order an ANA?

If autoimmune rheumatic disease is likely, the ANA can be helpful for diagnosis and classification.

If autoimmune rheumatic disease is unlikely, do not order an ANA. A positive ANA may cause anxiety, unnecessary investigations and potential confusion for both patients and providers.

Labs to consider in patients with non-specific symptoms and low probability of lupus

LOOK FOR EVIDENCE OF KIDNEY DISEASE

- Urinalysis, urine protein / creatinine ratio.
- Can be done conveniently on random 'spot' urine.

LOOK FOR (HEMOLYTIC) ANEMIA, THROMBO-CYTOPENIA, LYMPHOPENIA, NEUTROPENIA

- CBC with differential white count.

LOOK FOR RENAL FUNCTION, ACIDOSIS, LIVER DISEASE, MUSCLE INFLAMMATION

- Often but not always low in active disease.
- Acute phase reactants: elevated in other causes of inflammation (infection).

THYROTROPIN HORMONE (TSH)

- Fatigue, various symptoms in hypothyroidism and hyperthyroidism.

ASK ABOUT THROMBOSIS OR POSSIBLE ANTIPHOSPHOLIP ID SYNDROME

Lupus and mortality

Cardiovascular disease is the major cause of death in people with longstanding lupus.[9]

Factors contributing to increased mortality:

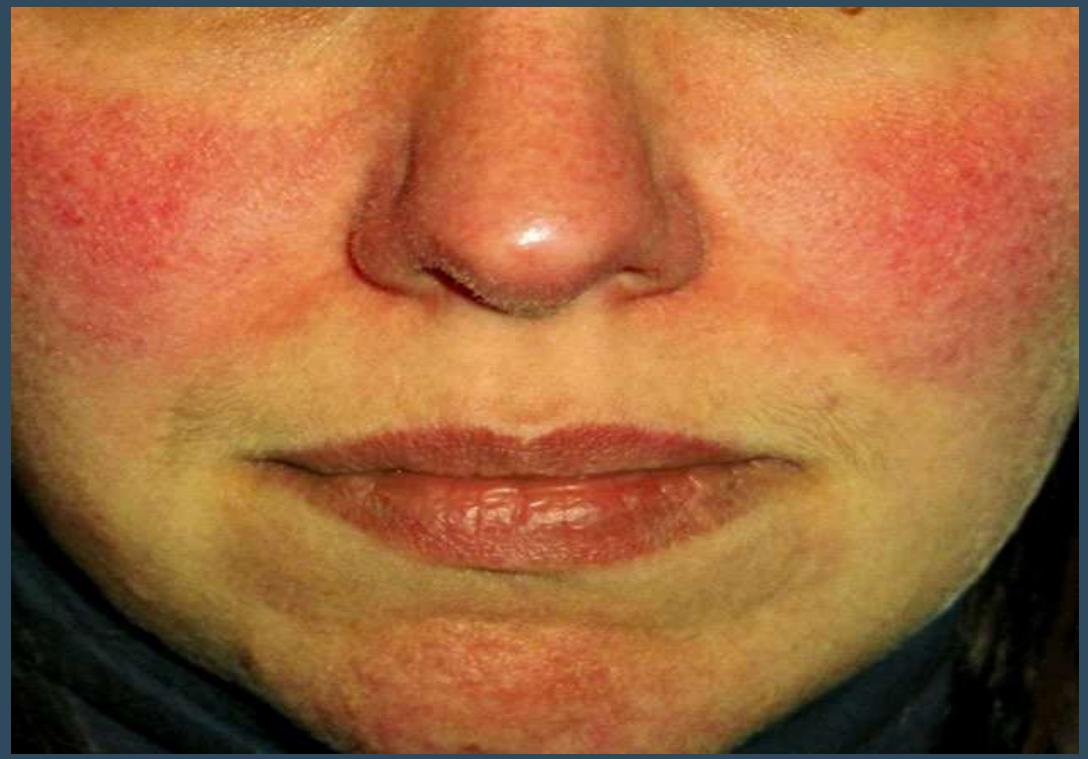
- Active lupus and infection
- High disease severity at diagnosis
- Younger age at diagnosis
- Ethnicity (Black/African American, Hispanic / Latino, Asian and Native American/Alaska Native)
- Male gender
- Low socioeconomic status
- Poor adherence to treatment protocol
- Inadequate support system
- Limited education

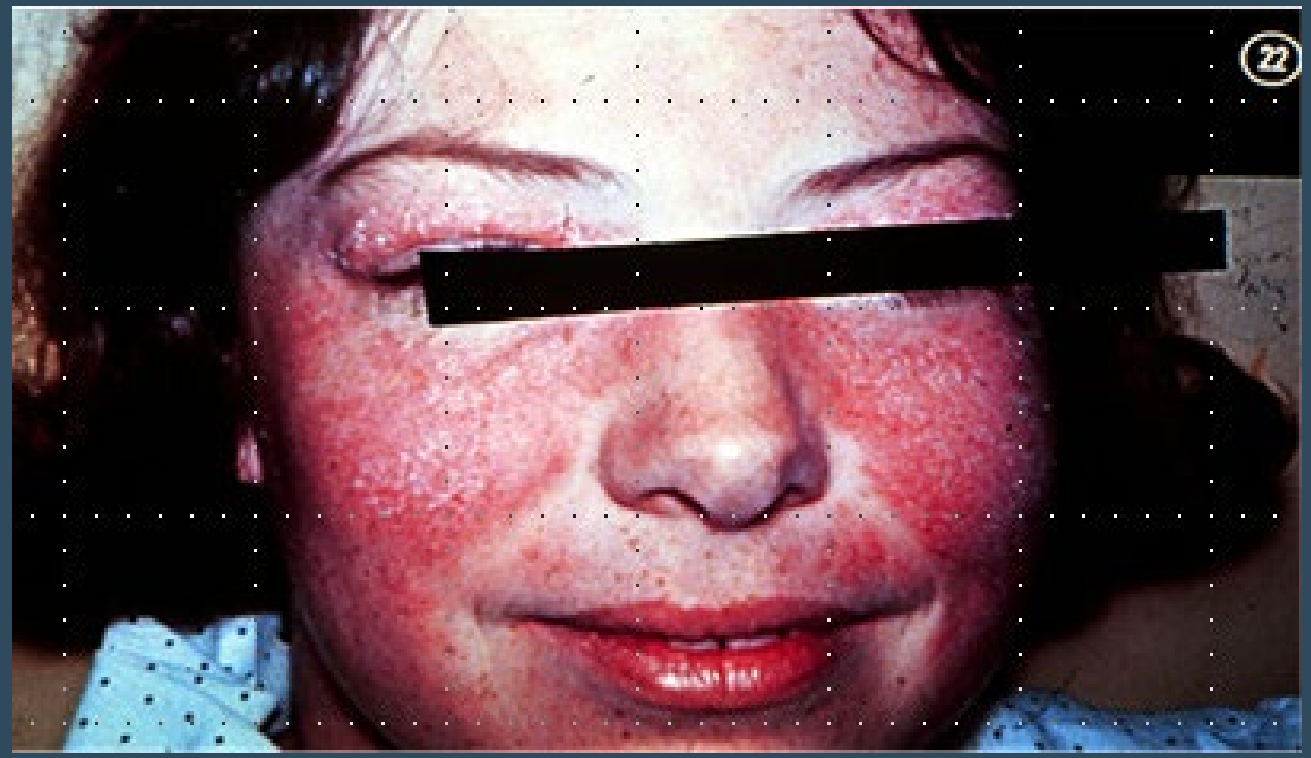
Importance of early referral

- Mortality is higher in people with lupus compared to the general population.
- Five-year survival rate in 1953 was 50 percent. Today the survival rate is 90 percent because of better detection and treatment.
- Currently 80 to 90 percent of people with lupus survive 10 years after diagnosis, but that drops to 60 percent with advanced stages of organ deterioration.
- The leading causes of mortality are preventable with appropriate therapies.[10]

Let’s look at some cases.

Which patient has lupus?





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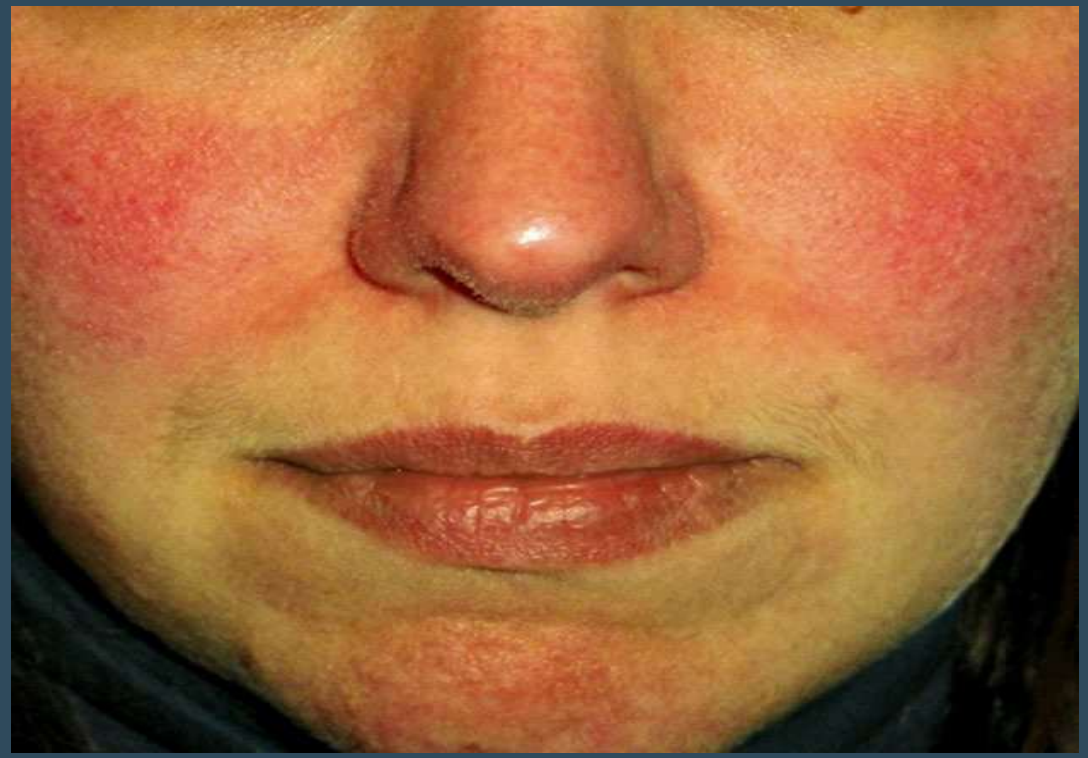

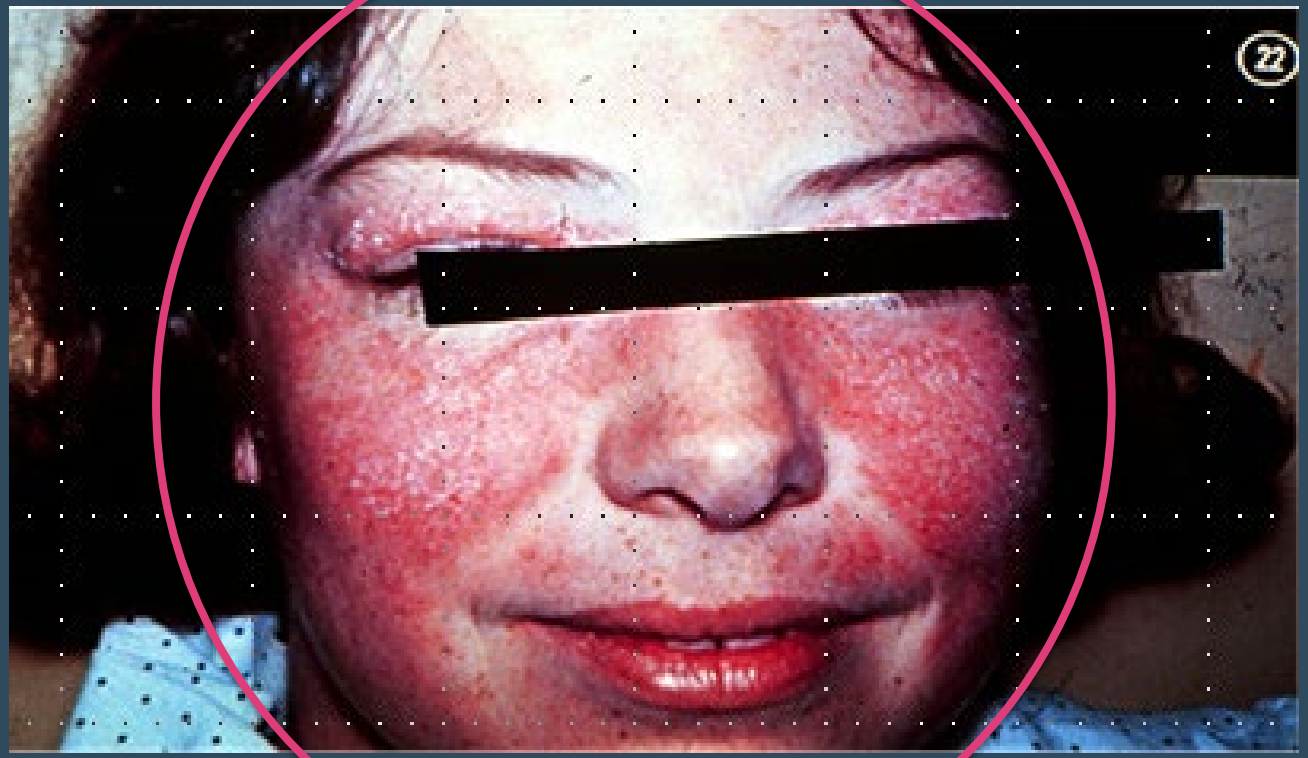

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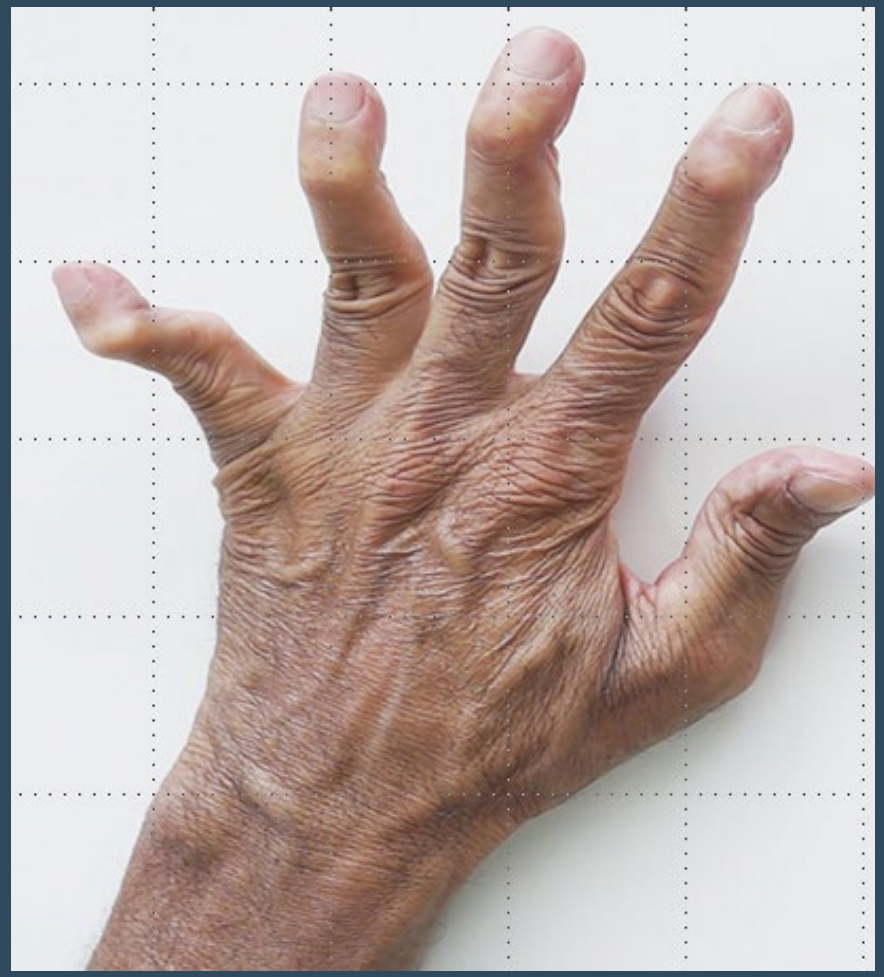

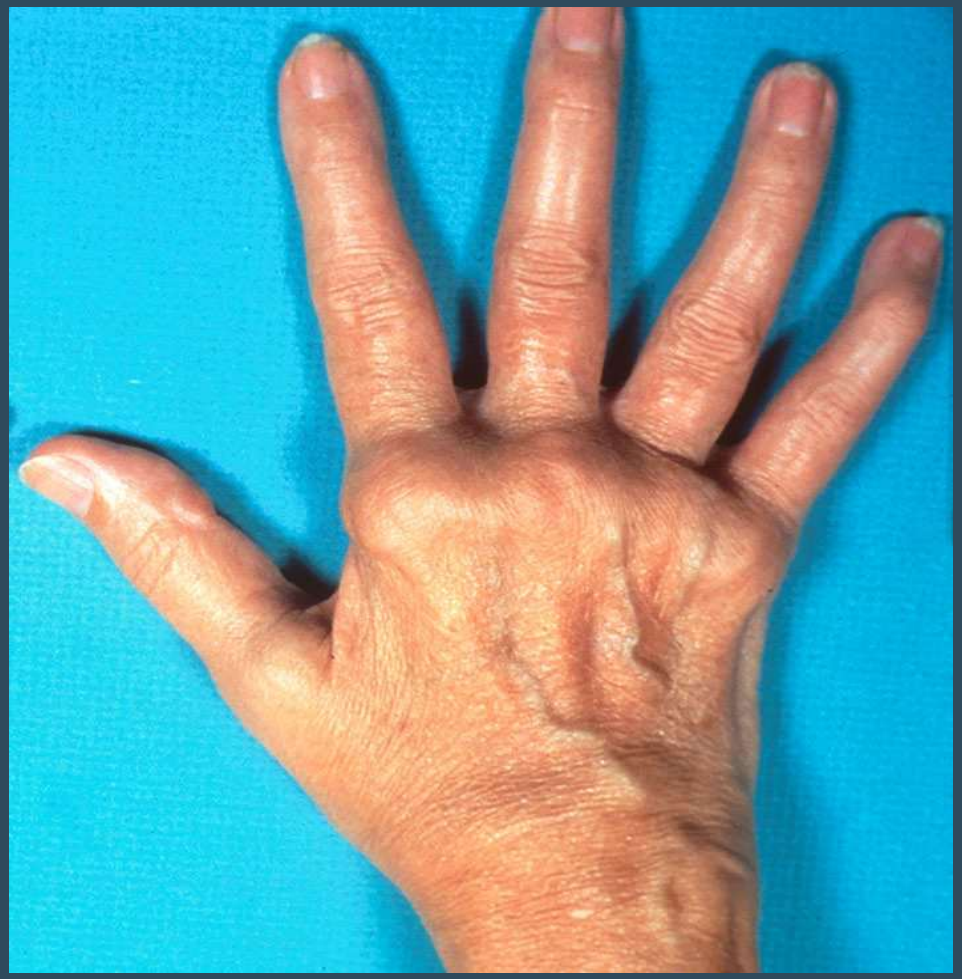

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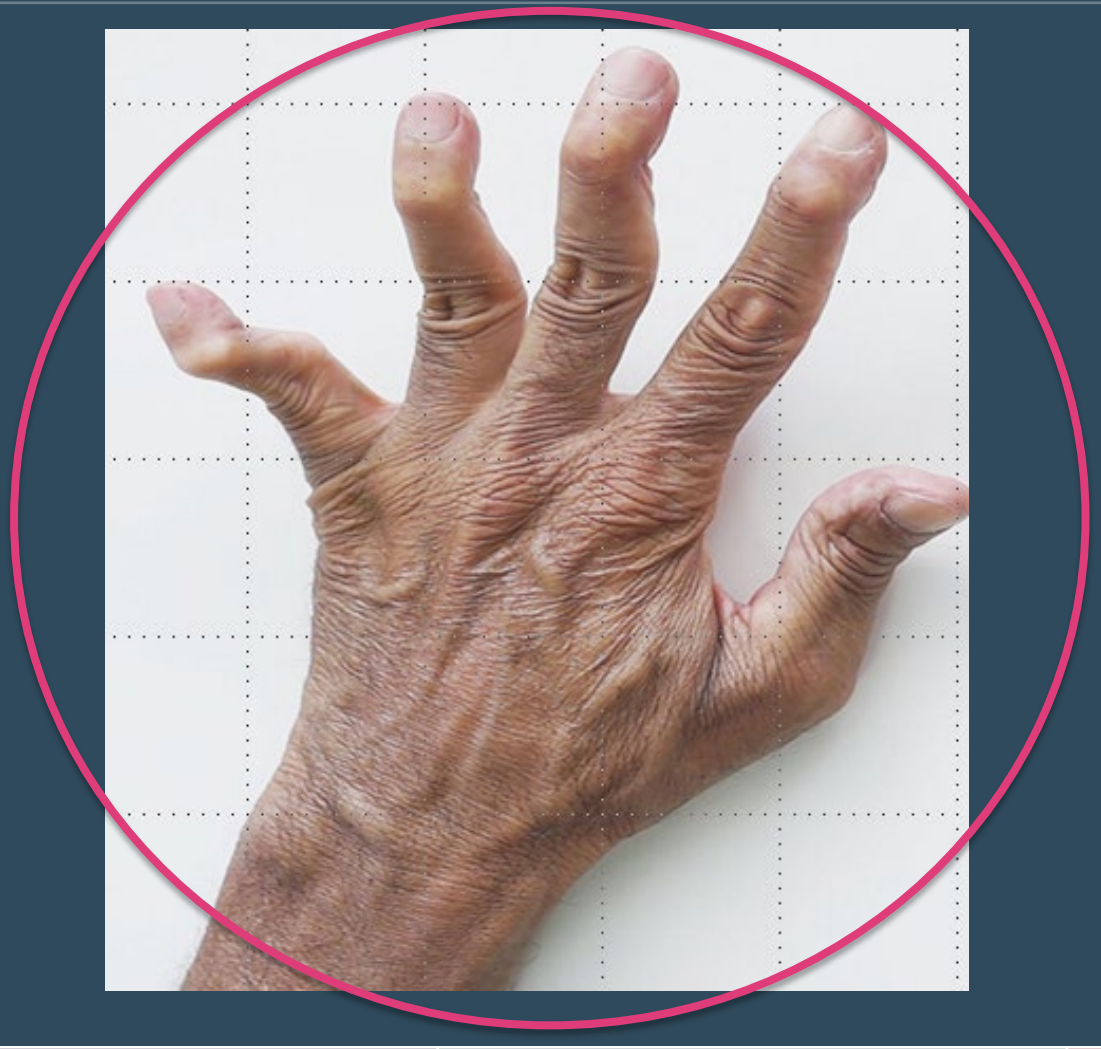

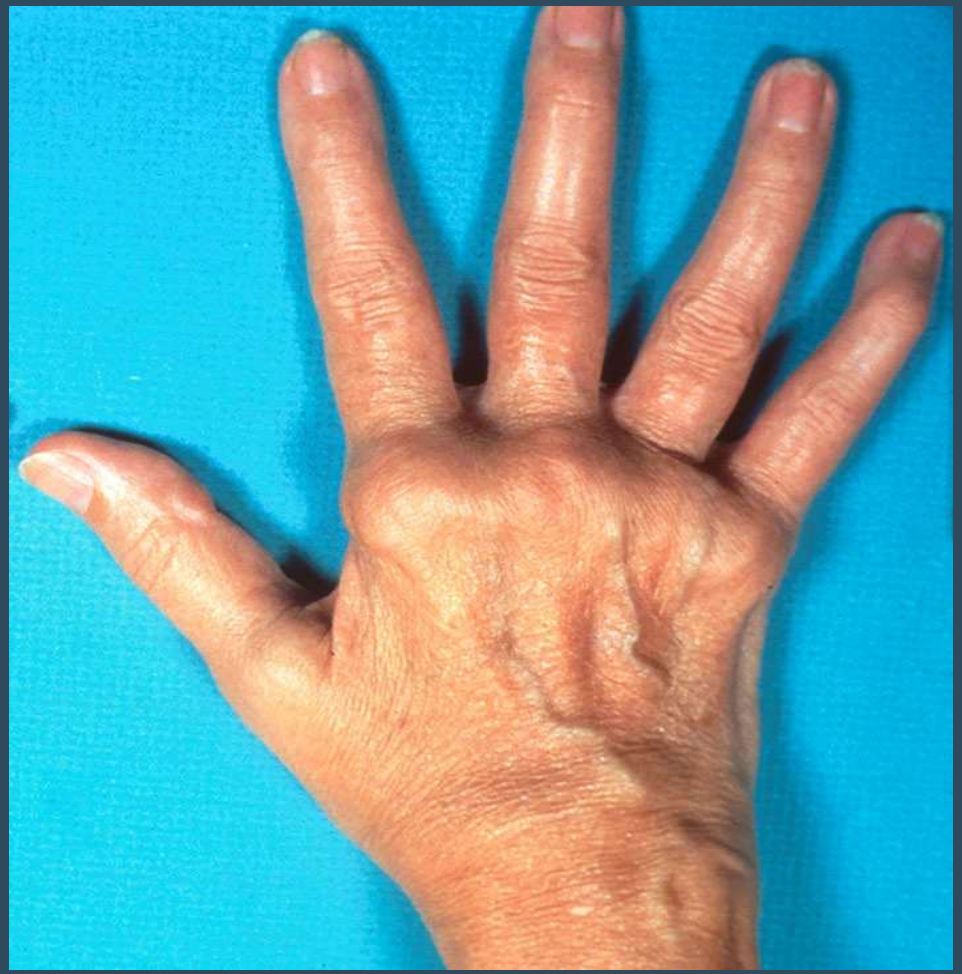

Which patient has lupus?

- 28 year old woman presents with fatigue and diffuse joint pain.
- She has had joint pain for 10 years but it is much worse now.
- She has migraines but no other symptoms.
- Exam shows no inflammation of joints, but she has multiple tender points.

- 28 year old woman presents with fatigue and diffuse joint pain.
- Her joint pain started about 4 months ago and has become worse.
- She has noted some new sores in her mouth and has had a rash with sun exposure recently.
- Exam shows swelling at PIP joints of hands.

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Case 1

- 39 year old woman has had swollen and stiff hand joints for one year (MCP and PIPs), with morning stiffness, referred to rheumatology for suspected rheumatoid arthritis
- ROS: photosensitivity
- Exam: patchy alopecia, inflammatory arthritis
- Labs: WBC 3.0, ALC 0.8, SCr 0.6, UA normal, ANA+ 1:320 titer, RF-, CCP-, Sm+, dsDNA-, SSA/SSB-, RNP-

- What is the diagnosis, and **why?**

Case 1

- 39 year old woman has had swollen and stiff hand joints for one year (MCP and PIPs), with morning stiffness, referred for suspected RA
- ROS: photosensitivity
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- What is the diagnosis, and **why?**

  SLE: +ANA, hematologic (leukopenia), arthritis, immunologic (Sm), and photosensitivity

Case 2

- 33 year old black woman presents with:
- Inflammatory arthritis, +ANA for 4 years
- 1 year ago: pericardial and pleural effusions, resolved with prednisone
- Now: fever, cough. Labs: SCr 3.8, WBC 3.4, C3 61↓, C4 9↓, 3.1 g proteinuria, UA lrg bld
- Autoantibodies: ANA/dsDNA/SSA/SSB/RNP+
- Other sx's: photosensitivity, Raynaud's, pruritic rash, oral ulcers

- What is the diagnosis, and **why?**

Case 2

- 33 year old black woman presents with:
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- Autoantibodies: ANA/dsDNA/SSA/SSB/RNP+
- Other sxs: photosensitivity, Raynaud's, pruritic rash, oral ulcers

- What is the diagnosis, and **why?**

  SLE: +ANA, hematologic (leukopenia), arthritis, immunologic (dsDNA), photosensitivity, mucosal ulcers, renal disorder (proteinuria), serositis, low complements

Case 3

- 47 year old white woman comes in wanting a lupus test. She has read about lupus and is convinced she has it.
- Sxs: aches all over her body and feels tired all the time. Has been present for a while but worse over past year. Has trouble sleeping and does not feel rested in AM.
- PMH: migraine headaches
- PE: no swollen joints, no rashes, no oral ulcers, multiple soft tissue tender points

Should you order an ANA now?

Case 3

- 47 year old white woman comes in wanting a lupus test. She has read about lupus and is convinced she has it.
- Sxs: aches all over her body and feels tired all the time. Has been present for a while but worse over past year. Has trouble sleeping and does not feel rested in AM.
- PMH: migraine headaches
- PE: no swollen joints, no rashes, no oral ulcers, multiple soft tissue tender points

Should you order an ANA now?

Neither ANA nor RF should be ordered without a strong clinical suspicion of disease. Initial work-up of fatigue and myalgia includes general labs, hepatitis C testing in appropriate age or risk groups, and thyroid function.

In summary

Early symptoms can be non-specific and be easily mistaken for other illnesses or syndromes.

Symptoms may be transient or prolonged and independent of one another.

Consider lupus if the person you are treating presents with vague complaints from the signs and symptoms list.

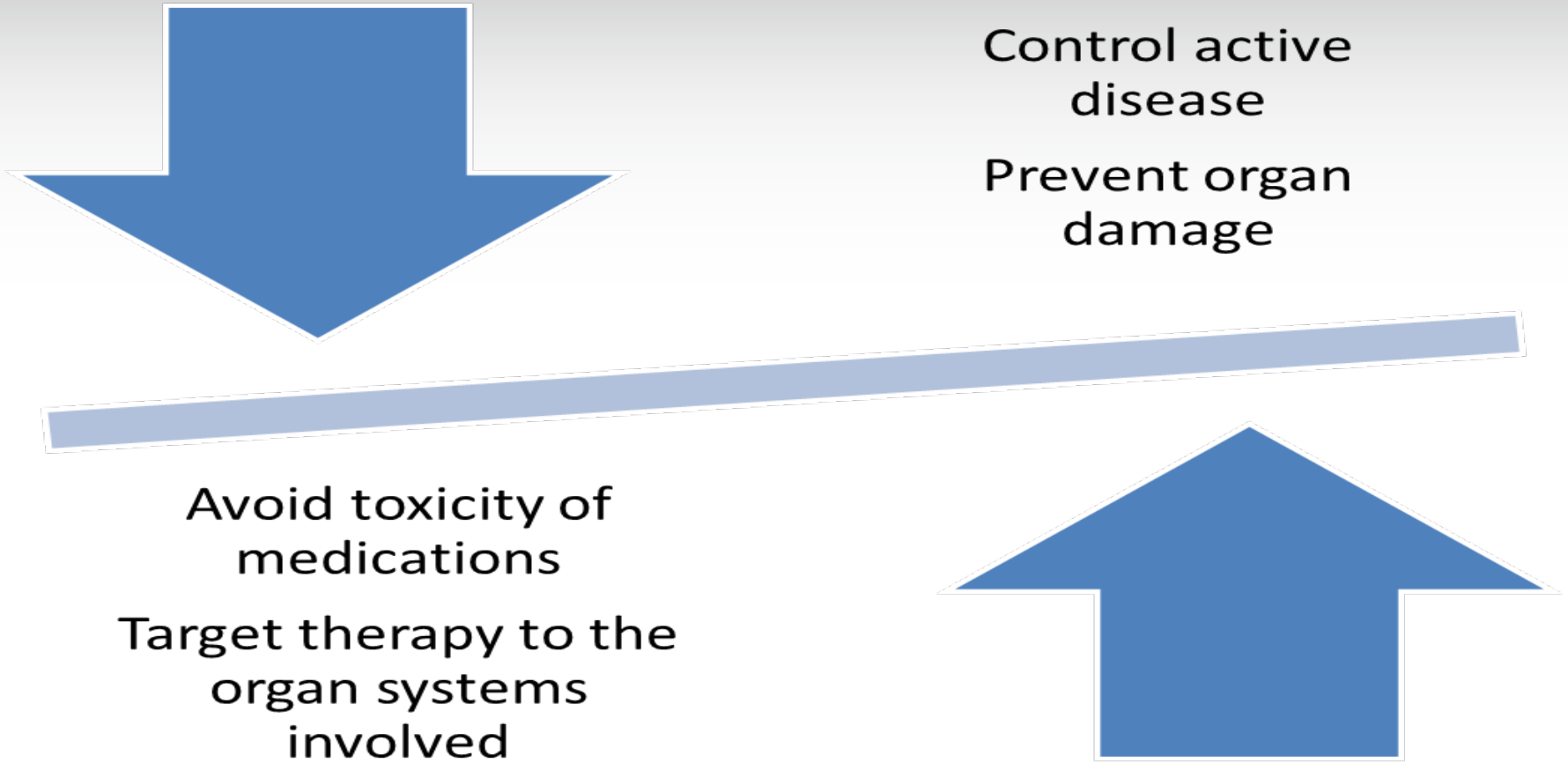
Also consider lupus if the person has a family history of autoimmune disease.

Do an initial screening, including: CBC, BMP, LFTs, ESR, CRP, ANA, UA

Refer to a rheumatologist for assessment and diagnosis.

Medications in Lupus

Goals of Therapy in SLE



Disease Activity

- Important determinant of need for ongoing immunosuppressive therapy
- Defined by lupus manifestations present NOW
- In contrast to “Damage” or organ dysfunction that has accumulated over time due to previous SLE disease activity

Disease Activity vs. Damage

There are 2 major types of symptoms from lupus, each treated differently.

Activity vs. Damage

NO LUPUS

The forest is your body. The fire is your immune system. Here, the fire is used for good.

LUPUS ACTIVITY

The fire (immune system) is attacking the forest (the body).

TREATING A FLARE

The water is the lupus medications.

LUPUS DAMAGE

After many years of flares, damage remains. It is a problem. It was caused by the fire. However, now, it does not respond to more water.

RECOVERY FROM DAMAGE

Recovery from damage may be slow and incomplete… but there is hope!

Disease Activity vs. Damage

There are 2 major types of symptoms from lupus, each treated differently.

- This may help you understand why your patients' symptoms may not completely go away over time and why medications that were previously helpful are not so much any more.
- This may explain why some of your patients often get told "your labs are fine", even though they just got through explaining how horribly they feel.
- Whether activity or damage, these are important and real issues your medical team needs to address. But they are treated differently.

Medications for SLE

- No drugs FDA approved from 1958-2010
- Only 4 drugs FDA approved
  - Aspirin
  - Corticosteroids
  - Hydroxychloroquine (Plaquenil)
  - Belimumab (Benlysta)
- Off-label use is common
  - Azathioprine, mycophenolate mofetil, cyclophosphamide, methotrexate, etc.

Corticosteroid Side Effects

- Short vs. long-term effects
  - Appetite, sleep and mood often affected short-term, particularly with higher doses
  - May raise BP and glucose in short-term
  - Longer-term effects of osteoporosis, accelerated CVD, and cataract formation
- Dose effect
  - The lower the dose, the less likely
  - Infection risk remains high even on lower doses

Complications of Corticosteroids

- Infections
- Weight gain
- Cushingoid appearance
- Osteoporosis
- Osteonecrosis
- Impaired glucose tolerance
- Mood disturbances
- Cataracts and/or glaucoma
- Increased risk of GI bleed if on NSAIDs
- Hypertension
- Lipid abnormalities
- **Premature atherosclerosis**
- Skin fragility/acne
- Myopathy
- Tendon rupture
- Withdrawal syndrome if not tapered
- Adrenal insufficiency

Corticosteroid Doses

- High Doses
  - Pulse dosing can be used early in severe disease
  - Must taper, amount of taper is unique to each patient and situation and should be tailored
  - Consider instituting a steroid-sparing agent
- Rarely use more than 10-15 mg/day on a chronic basis in non-organ-threatening, active disease
- For active arthritis, 5-10 mg per day

Anti-Malarials: First-line Therapy in SLE

- Hydroxychloroquine is the most commonly prescribed anti-malarial medication.
- Chloroquine is available but more toxic.
- Quinacrine may be used in some cases, but availability is limited.

Anti-Malarial History and Mechanism

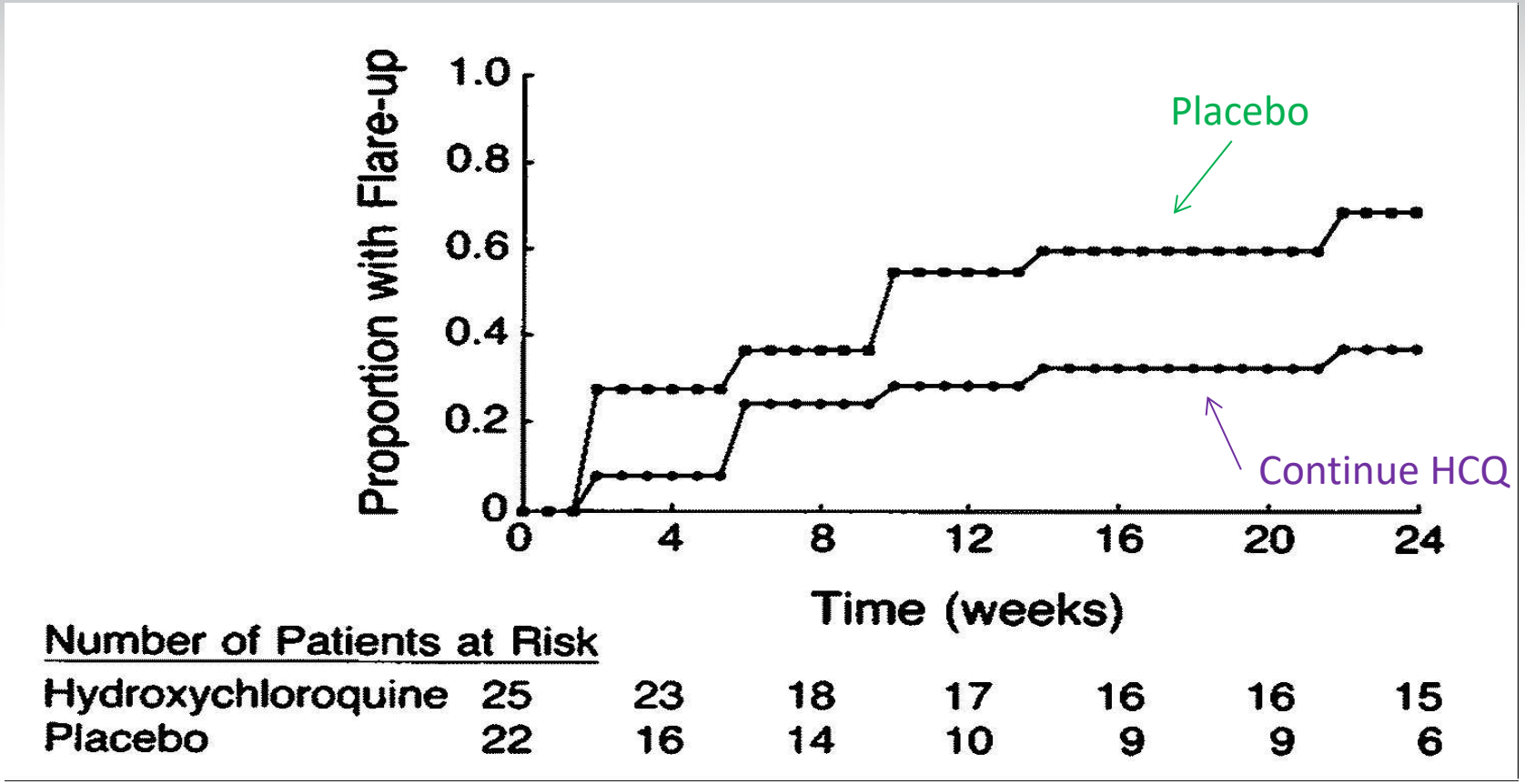
- Derived from bark of cinchona tree
- Analogous to quinine
- First used in SLE in 1894
- Raises intracellular pH
  - Inhibiting acid-dependent cellular processes such as protein processing
  - Turns off receptor activation sites
- Prevent activation of TLR 7 and 9
- Long half-life

Benefits of Hydroxychloroquine[11]

- Controls skin and joint disease
- Long-term use prevents major renal or CNS damage
- Protective effect on survival in SLE
- Lower fasting glucose in women with SLE or RA taking hydroxychloroquine
- Mild anti-thrombotic properties
- Favorable effect on lipids
- May have a role in preventing congenital heart block. This is not an infection.

Time to Flare: Continue HCQ vs. Withdraw[12]

Life Table of Time to a Clinical Flare-up for Patients Randomly Assigned to Continue Taking Hydroxychloroquine (Circles) or to Receive Placebo (Squares).



the NEW ENGLAND JOURNAL of MEDICINE

The Canadian Hydroxychloroquine Study Group*. N Engl J Med 1991;324:150-154.

Toxicity of Hydroxychloroquine

- **Safe in pregnancy**
- No blood tests needed for monitoring
- Is not immunosuppressive
- Does not impair wound healing
- Retinal toxicity, though very rare, is the biggest concern
  - Risk increases with cumulative lifetime dose
- Other possible side effects:
  - GI, skin, neuromuscular, cardiomyopathy, CNS

Azathioprine (Imuran)

- **Can be used in pregnancy**
- Useful in maintenance for lupus nephritis, other severe manifestations, steroid-sparing
- Toxicity:
  - Bone marrow suppression, hypersensitivity reaction, nausea/vomiting, ↑LFTs
  - TPMT enzyme activity assessment prior to start helps identify the few people with no enzyme activity

Mycophenolate Mofetil (CellCept)

- **Not recommended in pregnancy (Category D)**
- Targets lymphocytes preferentially
- Useful in lupus nephritis, lung disease, and other severe manifestations
- Toxicity:
  - GI (N/V/D), leukopenia, anemia

Cyclophosphamide

- Originally used as chemotherapy
- Has immunosuppressive effects useful for autoimmune diseases
- Two IV treatment regimens
  - Low dose every two weeks for several months
  - Higher dose monthly for 3-6 doses
    - is the most common regimen used for induction therapy for lupus nephritis or CNS lupus
- Can be given oral, but IV less toxic
- CBC prior to dose and 10-14 days after; regular UA with microscopy
- Toxicity:
  - bone marrow suppression, N/V, alopecia, **teratogenicity,** gonadal dysfunction, hemorrhagic cystitis, bladder cancer, lymphoma, infections

Methotrexate

- Most effective in arthritis of SLE
- Not effective for internal organ involvement
- Dosed as in RA (up to 20mg PO/25 mg SQ weekly)
- Given with folic acid 1 mg/d
- Toxicity:
  - Bone marrow suppression, GI, alopecia, photosensitivity, **teratogenicity**

Complications of Immunosuppressants and Cytotoxics

- Infection
- Cancer
- Infertility
- Teratogenicity

Biologics

Definition:

- A substance that is made from a living organism or its products and is used in the prevention, diagnosis, or treatment of disease in humans. Biological drugs include antibodies, interleukins, and vaccines.

Belimumab (Benlysta)

- FDA approved for treatment of SLE in 2011
- Monoclonal antibody vs. BLyS (B-lymphocyte stimulator)
- 2 trials for FDA approval added belimumab to standard of care in active SLE
  - Excluded severe SLE such as nephritis
- 10 mg/kg IV at 0, 2, and 4 weeks, then every 4 week infusion
- ~$2000 per dose
- Role in care for SLE still being determined

Medication Access

- Hydroxychloroquine
  - Generic, 60 tabs $80-110
- Immunosuppressants
  - Mycophenolate 120 tabs of 500 mg: $55 generic, ~$1800 for brand name
  - Azathioprine ~$25 for 60 tabs of 50mg
  - Methotrexate ~$40 for 24 tabs of 2.5mg
  - Cyclophosphamide usually given IV
- Biologics
  - Very expensive!

Non-Pharmacologic Interventions

- UV protection
- Discuss avoidance of sulfonamide-containing antibiotics
  - May need to weigh benefits and risks
- Avoidance of immunizations with live virus
- Smoking cessation
- Standard immunizations
- Monitor lipids, BP, bone density
- Birth control
- Incorporate exercise or physical activity and diet as part of your treatment under the advice of your healthcare professional

Vaccination of Immunocompromised Persons

Some vaccines specifically indicated for immunocompromised

- Pneumococcal conjugate 13 valent vaccine (PCV13)
- Pneumovax
- Influenza

Other vaccines specifically contraindicated in immunocompromised

- Live vaccines
  - MMR
  - Varicella
  - Zoster
  - FluMist

Case 1

- 19 year old woman with new diagnosis of lupus with nephritis
- Presented with anasarca and elevated BP
- Other findings: leukopenia, lymphopenia, Coombs+ anemia, thrombocytopenia
- Nephrotic range proteinuria; hematuria
- +ANA, +dsDNA, low C3 and C4
- Diagnosed with class IV lupus nephritis (diffuse proliferative glomerulonephritis)

Case 1

- Lupus nephritis in a 19 year old woman
- Aggressive Rx in short term
- Long term goals related to lupus:
  - Reduce corticosteroid exposure
  - Prevent damage

Recommendations in this Case

- Appropriate nephritis therapy
  - Corticosteroids and immunosuppressive agent
  - Aim to taper off steroids
- Blood pressure control
- ACE or ARB if proteinuria
- Use of contraception

Case 2

- 51 year old man presented with arthralgias for 3 years presents for rheumatology evaluation
- No other symptoms suggesting other organ system involvement
- PE: bilateral 2nd/3rd MCP joint swelling in the hands
- The rest of the exam was unremarkable.

Case 2 Continued

- Labs:
  - RF and CCP negative
  - ANA+, dual pattern, 1:320 speckled and 1:40 homogeneous
  - Anti-dsDNA positive, SSA/SSB positive, Sm and RNP negative

What medication do you recommend for this patient?

Case 2 Continued

- Started on hydroxychloroquine and joint symptoms and swelling improved
- Flare of arthritis had occurred and he was treated with a prednisone taper, starting at 20 mg daily
- Repeat history and exam: no signs or symptoms to suggest other organ system involvement
- Repeat labs: dsDNA remains positive, WBC 4.9, Abs. lymphocyte count 0.9, no anemia or thrombocytopenia, C3 and C4 normal, UA unremarkable, serum creatinine 0.8

What do you think of the prednisone dose?

Would you change anything in his regimen now?

Case 2 Discussion

- SLE diagnosis based on:
  - Positive ANA
  - Positive anti-dsDNA antibody
  - Lymphopenia
  - Arthritis
- Mild disease, joint signs/symptoms and laboratory abnormalities only
- Treatment with hydroxychloroquine has been continued and he continues to do well without any recurrent flares

Summary

- SLE is a heterogeneous disease and treatment depends on organs involved and severity.
- Goal of minimizing medication toxicity applies especially when considering corticosteroids.
- Understanding the pathogenesis of SLE is leading to development of more targeted Rx
- **Don’t forget Plaquenil (hydroxychloroquine)!!**

Health maintenance and preventive care considerations in lupus.

Why is health maintenance and preventive care in patients with SLE so important?[15]

- SLE patients are at high risk for comorbidities and complications (CVD, infections, Osteoporosis) that can be mitigated by appropriate preventive care
- However, the provision of preventive care in patients with lupus is suboptimal
- Proportion of SLE patients who receive recommended Primary Care Services:
  - → Influenza vaccine: 57%
  - → Pneumonia vaccine: 49%
  - → Cholesterol Monitoring: 65%
  - → Low dose aspirin: 52%
- SLE patients who are less likely to receive primary preventive services include:
  - → Younger patients
  - → Uninsured
  - → Individuals who do not have a PCP

Learning objectives related to health maintenance and preventive care:

- Know the risk factors for premature coronary artery disease in lupus patients and risk modification strategies
- Recognize osteoporosis risk factors and its treatment considerations in women of childbearing age
- Identify appropriate immunization schedules for lupus patients and understand contraindications to immunization among patients on immunosuppressive medications

ACE Inhibitors for Nephritis & Proteinuria[16]

IF a patient with SLE has renal disease (**proteinuria** >=300 mg/day or **eGFR <60** ml/minute) **and** 2 BP readings, including the last reading, with **systolic BP > 130** mm Hg or **diastolic BP > 80** mm Hg over 3 months

THEN pharmacologic **therapy for hypertension** should be initiated or the current regimen should be changed or escalated, unless patient refusal or contraindications are noted.

IF a patient with SLE has proteinuria 300 mg/day

THEN the patient should be treated with an ACE inhibitor or ARB, unless patient refusal or contraindications are noted.

Teratogenic Drug Counseling for Reproductive Age Women[16]

IF a woman between 18 and 45 years of age is started on any of the following medications for SLE: chloroquine, quinacrine, methotrexate, azathioprine, leflunomide, mycophenolate mofetil, cyclosporine, cyclophosphamide, or thalidomide

THEN a discussion with the patient about the potential teratogenic risks of therapy and about contraception should be documented prior to drug initiation, unless the patient is unable to conceive (e.g., has had a hysterectomy, oophorectomy, tubal ligation, or is postmenopausal).

Cardiovascular Risk Assessment[16]

IF a patient has SLE

THEN risk factors for cardiovascular disease, including the following should be evaluated annually.

- smoking status
- BP
- BMI
- Diabetes
- serum lipids (including total cholesterol, HDL, LDL, and triglycerides)

Pneumococcal Vaccination[16]

IF a patient with SLE is on immunosuppressive therapy

THEN an inactivated influenza vaccination should be administered annually, unless patient refusal or contraindications are noted.

THEN a pneumococcal vaccine should be administered, unless patient refusal or contraindications are noted.

Bone Mineral Density Testing, Treatment in Steroid Users, Osteoporosis Treatment[16]

IF a patient with SLE has received prednisone (or other glucocorticoid equivalent) >=7.5 mg/day for >=3 months

→ THEN the patient should have BMD testing documented in the medical record, unless the patient is currently receiving antiresorptive or anabolic therapy.

IF a patient with SLE has received prednisone (or other glucocorticoid equivalent) >=7.5 mg/day for >=3 months

→ THEN supplemental calcium and vitamin D should be prescribed or recommended and documented.

IF a patient with SLE has received prednisone (or other glucocorticoid equivalent) >=7.5 mg/day for >=1 month, and has a central T score less than or equal to -2.5 or a history of fragility fracture

→ THEN the patient should be treated with an antiresorptive or anabolic agent, unless patient refusal or contraindications are noted.

If you need more information or have questions

Presenting rheumatologist, *please (1) delete this note and (2) insert the details for how and where to refer patients with suspected or diagnosed lupus and/or how they can contact you after the session.*

POST-TEST

IMPORTANT: A post-test survey will be emailed to you soon.

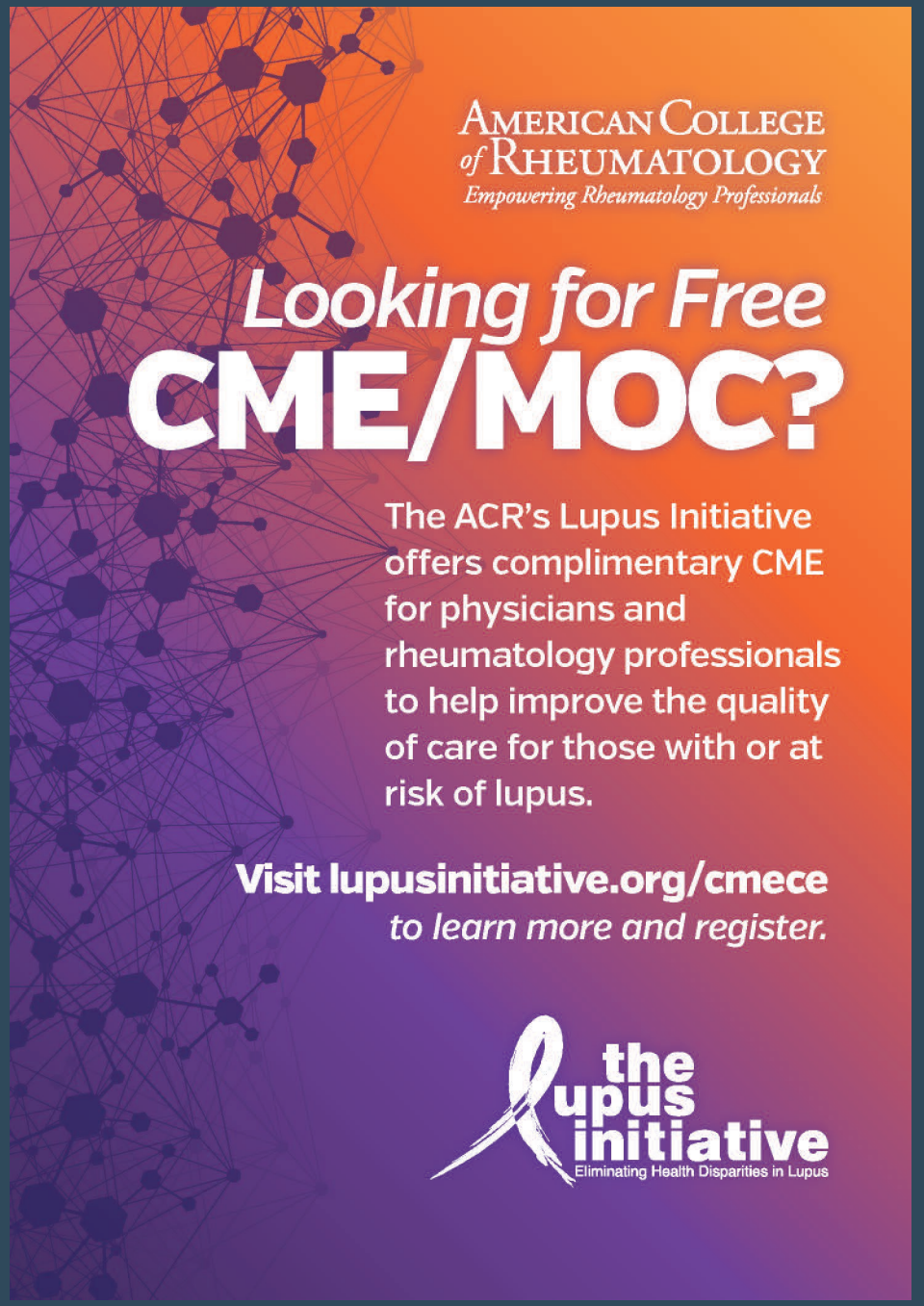
Please complete the post-test survey in order to receive your certificate.

Resources and Information

Ongoing care for people with lupus is a team effort, and it is a matter of life and death.

For presentations, videos, interactive case studies and CE/CME courses, visit The Lupus Initiative at https://thelupusinitiative.org/

For self-management resources, please visit: https://selfcare.thelupusinitiative.org/



Thank you.

This project is supported by the Centers for Disease Control and Prevention of the U.S. Department of Health and Human Services (HHS) as part of a financial assistance award totaling $7 million dollars with 100 percent funded by CDC/HHS. The contents are those of the author(s) and do not necessarily represent the official views of, nor an endorsement, by CDC/HHS, or the U.S. Government.

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