Different Types of Lupus…

SYSTEMIC
LUPUS
ERYTHEMATOSUS

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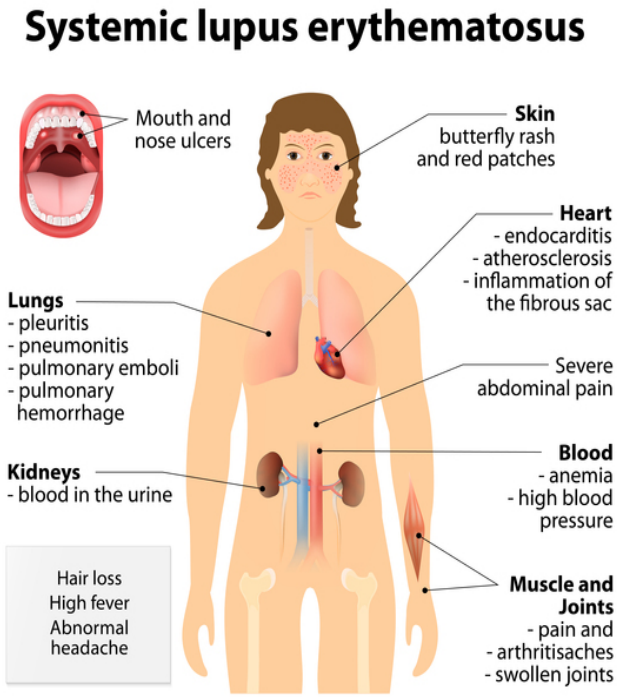
Rush Annual Lupus Educational Event

What is Lupus?

- SLE is a complex autoimmune disorder
- What is immune system?
- In Lupus immune system attacks own body's cell and tissue, resulting in inflammation and damage
- SLE can affect any part of the body, but most often harms the heart, joints, skin, lungs, blood vessels, liver, kidney, and nervous systems



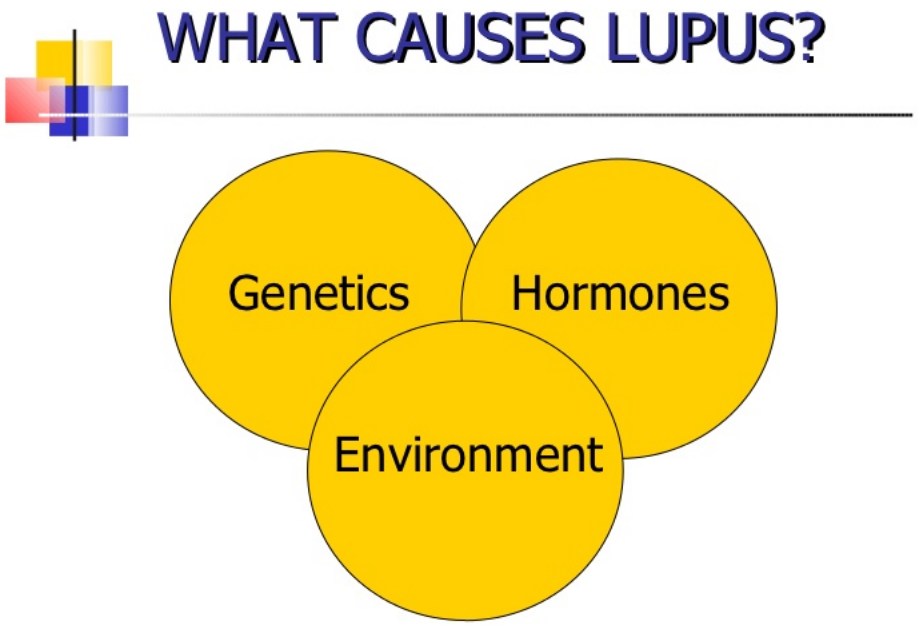
SLE

- Waxing and waning course
- Female to Male ratio of 9:1 (childbearing years)
- 70% of SLE: females between ages 15-45/ 10% present age >60
- Disease in males is can be more severe
- 1.5 million cases of lupus in USA
- Incidence 1.4-2.2 cases per 100,000 a year
- Prevalence 17 to 48 per 100,000 population

Lupus Facts

- Highest occurrence is in Afro-Caribbean females 1:250
- African American to Caucasian ratio 3:1
- Child of SLE mother - risk of SLE 1:15 (7%)
- 10-15% of SLE patients have 1st degree relative with SLE

What causes Lupus?



•Certain genes are more likely to occur in patients with lupus

- Many of these genes encode components of the immune system.
- Over 40 different genes predispose to SLE

•Environmental Factors: UVB light, chemicals, drugs, infections (Parvovirus, CMV, HCV), smoking

•Abnormal estrogen metabolism

- In animal studies estrogen worsens disease activity and causes early mortality

ANA

- ANA stands for “anti nuclear antibody”
- It is an important screening tool for diagnosis of SLE
- A positive ANA does NOT mean diagnosis of lupus
- Low Positive (1:160 or lower): SLE likelihood <2%
- Can be positive in many healthy people and other conditions (recent infection, other autoimmune diseases)
- ANA can be negative in the cutaneous lupus, especially discoid lupus

Specific Antibodies

- Anti-dsDNA: very specific, may correlate with disease activity
- Anti-Sm: specific, but only present in 25% of cases, does not correlate with activity
- APLA: not specific. Used to identify patients at increased risk for clots, thrombocytopenia and fetal loss

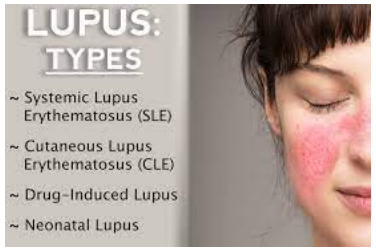
Criteria for the Diagnoses of SLE

- Malar (Butterfly) Rash
- Discoid Rash
- Sensitivity to the sun (Photosensitivity)
- Ulcers in the nose and mouth
- Arthritis
- Fluid around the heart, lungs and in the abdomen

- Lupus kidney disease
- Neurologic Disorders:
  - Stroke, inflammation, depression, memory dysfunction, etc…
- Anemia, low platelets and low white blood cell count
- Abnormal blood antibody levels
- ANA blood test

Lupus Types…

- Systemic Lupus- SLE
- Cutaneous Lupus- CLE
- Drug Induced Lupus
- Neonatal Lupus



DRUG INDUCED

- Drug Induced SLE
- Develop after the patient taking a known lupus inducing drug for at least 1 month (usually months to years) - (Bactrim, Minocycline, HCTZ, Hydralazine, PTU)
- Positive ANA and anti-histone
- Clinical features: arthritis, myalgia, rash, fever, serositis, splenomegaly

Drug Induced SCLE (DI-SCLE)

- DI-SCLE first described with HCTZ in 1985.
- > 40 drugs associated with SCLE (ACE inhibitors, beta blockers, calcium channel blockers, terbinafine).
- Incubation period days to years.
- Clinically/pathologically identical to idiopathic SCLE.
- ANA and anti-SSA common, often persist.
- Anti-histone usually absent.
- Resolves within weeks of cessation of the drug.
- DI-SCLE is distinct from DI-SLE.

Untitled

CLE

Cutaneous Lesions in Lupus

- **"Lupus specific" lesions-**
  characterized by interface dermatitis.
  - Acute cutaneous lupus (ACLE).
  - Subacute cutaneous lupus (SCLE).
  - Chronic cutaneous lupus (CCLE).
- **"Lupus non-specific" lesions.**
  - Livedo reticularis and livedo racemosa.
  - Retiform purpura.
  - Periungal erythema.
  - Leukocytoclastic vasculitis.

Skin

- Photosensitive
- Malar
- Discoid Lupus
- Subacute Cutaneous Lupus SCLE
- Alopecia Patchy
- Bullous
- Panniculitis

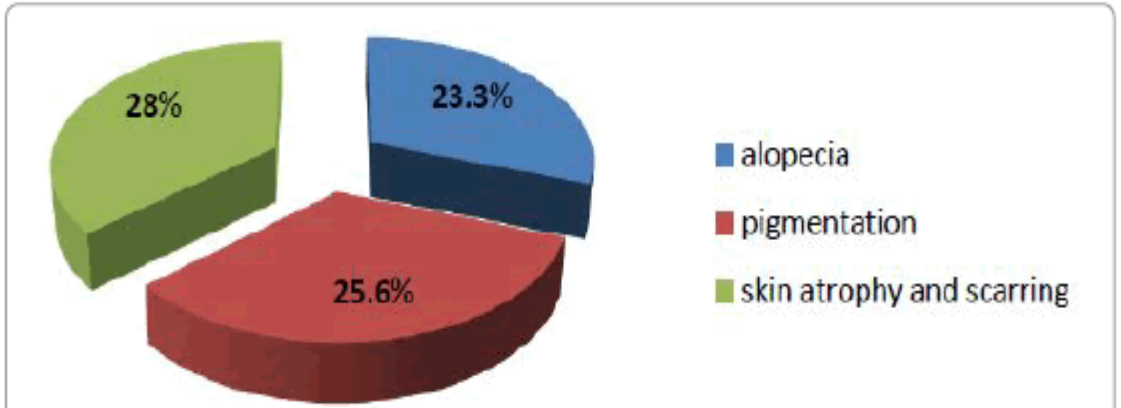


Figure 1: Outcome of cutaneous manifestation of systemic lupus erythematosus.

Photosensitivity

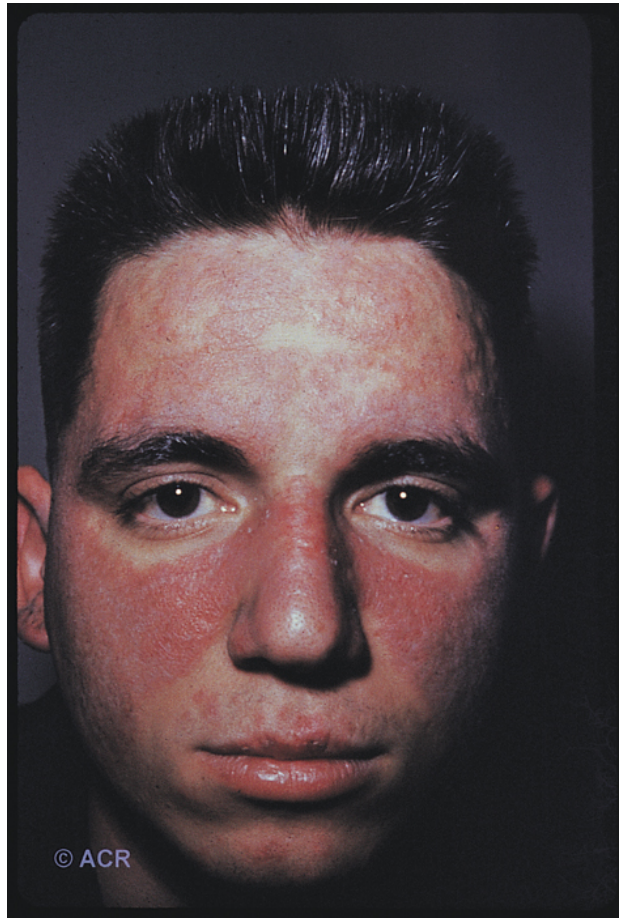
- Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation
- Acute or chronic
- Expected duration ~ 2 days
- Hives – seen in 10%/ pruritic

Butterfly rash

- Sounds kind of pretty when you toss the word butterfly in front of it, but it’s not cute.
- It’s red, it’s itchy, it takes over your face.
- It may cause others to stare, and for those suffering with it to want to shy away from going out in public.
- So until it subsides you are truly a butterfly warrior.

Malar (butterfly) Rash

- Fixed red, flat or raised, over the bridge of the nose and cheeks
- Tends to spare the nasolabial folds



Malar (butterfly) Rash

Discoid Rash

- Red raised patches with scaling
- Can be very scarring
- Can be lasting

Skin DLE

SCLE – subacute cutaneous

Panniculitis

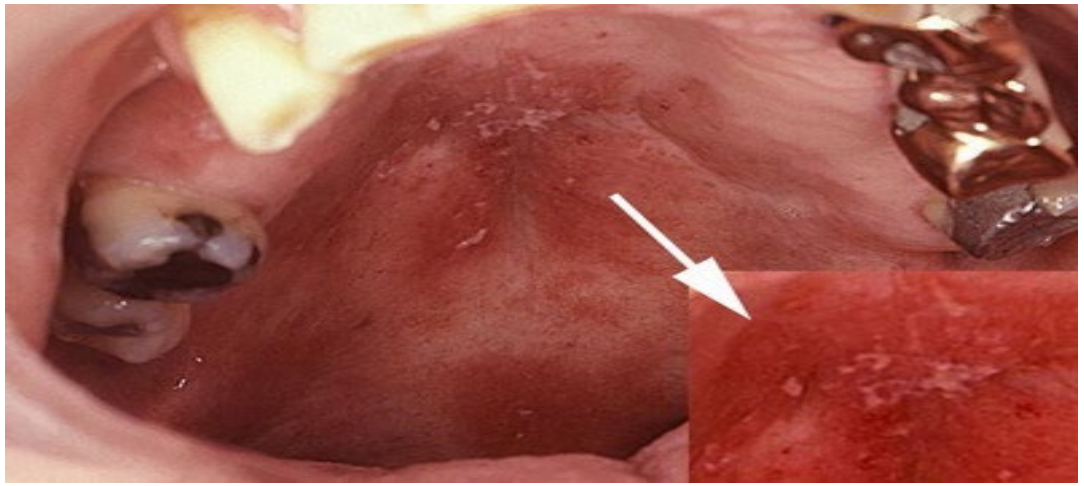
FIGURE 4: Lupus panniculitis - depressed erythematous violaceous nodules on the arms

Hair thinning

- Hair loss in patches- called alopecia
- Or just hair thinning / loss

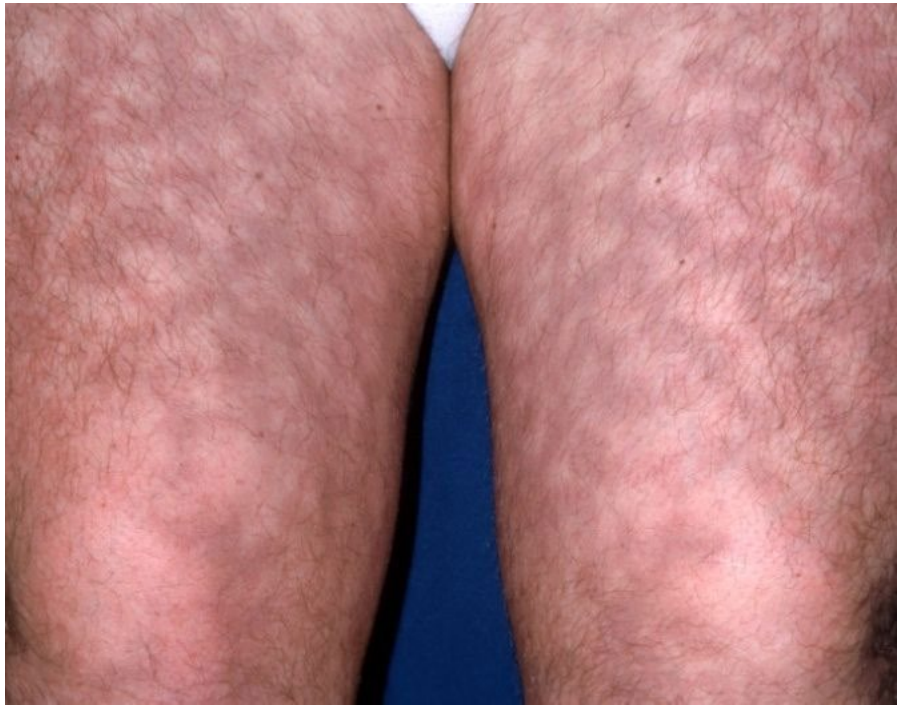
Oral and Nasal Ulcers

- Oral or nasopharyngeal ulcers
- Usually painless

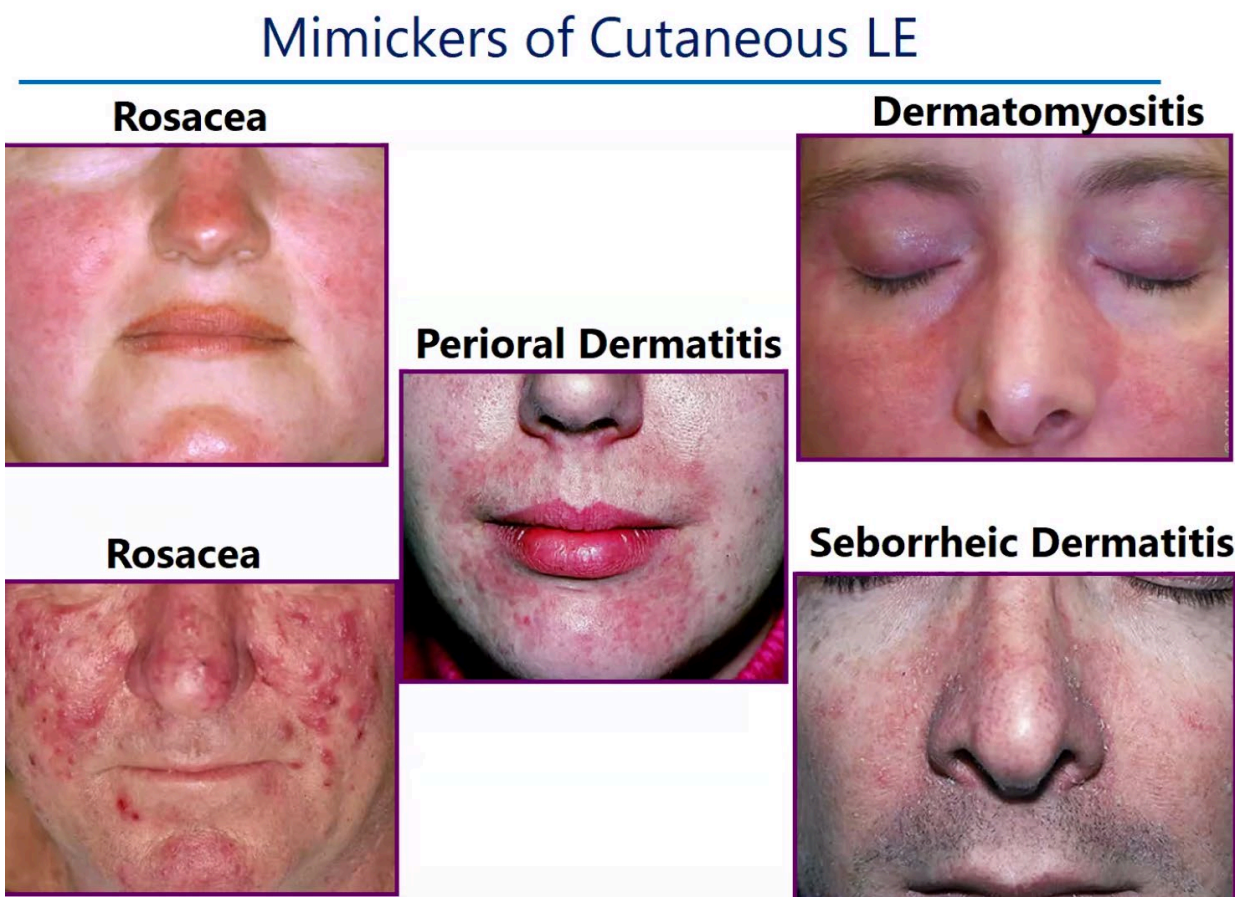


Raynaud’s Phenomenon

Livedo Reticularis



MIMICERS

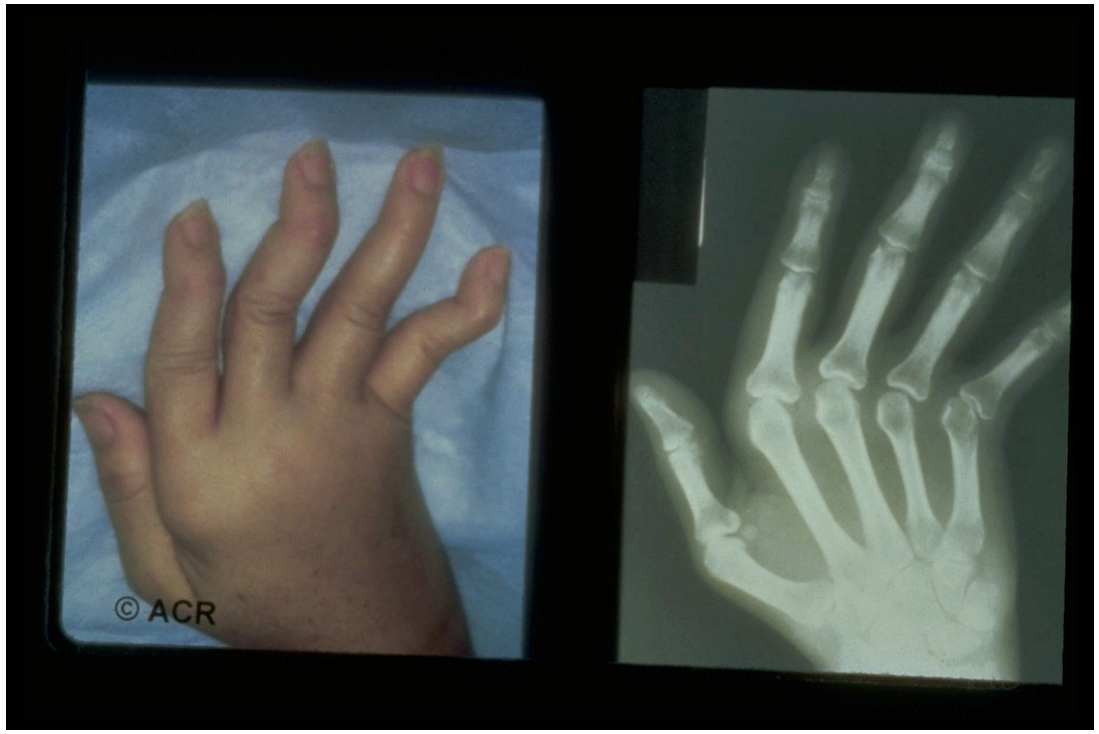

Mimickers of Cutaneous LE
Rosacea
Dermatomyositis
Perioral Dermatitis
Rosacea
Seborrheic Dermatitis

Musculoskeletal

- Arthritis vs Arthralgia?
- Hands, feet, knees, shoulders, hips
- Avascular necrosis (monoarticular- steroid use)

Arthritis vs Arthralgia

- Non-erosive arthritis involving two or more joints, characterized by pain, swelling, or fluid collections
- About 80% of patients have it
- Can move deformed fingers back into position- Jaccoud's arthropathy
- Joint deformities occur in only 10%

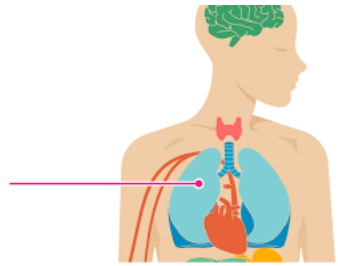


Serositis - Pulmonary

- Pleuritis – inflammation of the outer layer of the lungs
- With or without effusion (fluid)

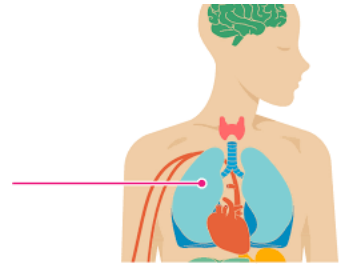
- Life-threatening manifestations: inflammation of lung itself which can lead to fibrosis/ ILD and intra-alveolar hemorrhage.

- Also pneumothorax and pulmonary HTN can occur



Cardiac

- Pericarditis: most common cardiac manifestation and usually responds to NSAIDs/ Pericardial effusions/ Tamponade
- Myocarditis (rare) and endocarditis (Libman-Sacks) may occur. Steroids plus treatment for CHF/arrhythmia or embolic events.
- MI due to atherosclerosis can occur in <35 y/o (steroids, lipids, smoking)



Neurological

- Cranial or peripheral neuropathy occurs in 10-15%
- Diffuse CNS dysfunction: memory and reasoning difficulty
- Headache: if excruciating, often indicate acute flare
- Seizures of any type
- Psychosis: must distinguish from steroid-induced psychosis
- Stroke/ transverse myelitis

Hematological

- Hematological abnormalities are frequent in SLE (66-86% of patients)

- Anemia: usually normocytic
- Leukopenia
- Thrombocytopenia

- APLA

Renal

- Lupus Nephritis: usually asymptomatic, so always checking UA if patient has known or suspected SLE
- Occurs early in course of the disease
- Histologic classification by renal biopsy is useful to plan therapy

What about Fatigue?

Fatigue

- Up to 80 percent of people with lupus experience fatigue
- Fatigue may be the main symptom and can be debilitating
- Contributing Factors:
  - Disease activity, pain, age, and medications
  - Poor physical and mental health
  - Lack of good social support
  - Smoking

Different SLE Types?

- Type I and II - recently proposed
- Validating patient perspective of the disease
- PRO- capturing a more comprehensive spectrum of symptoms

Symptoms

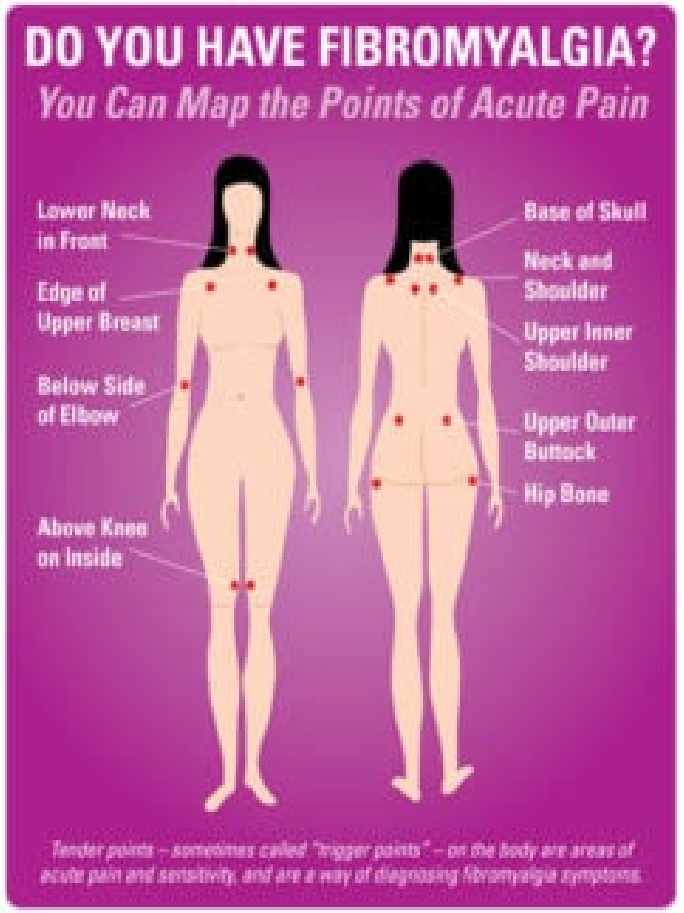
- Fatigue, and widespread pain represent the most common symptoms of SLE, despite not being part of classification criteria
- Etiology multifactorial (immunologic and nonimmunologic factors)
- Difficult to quantify, monitor or treat
- May not receive adequate attention by care team

How about Fibromyalgia?

- Regardless of SLE
- Higher pain level, fatigue
- Sleep dysfunction
- Mood disorders
- 20% vs 2-6% FM rates

*Intertwined and common features in both

Fibromyalgia - Researchers believe that fibromyalgia amplifies painful sensations by affecting the way your brain and spinal cord process painful and nonpainful signals.



SLE types

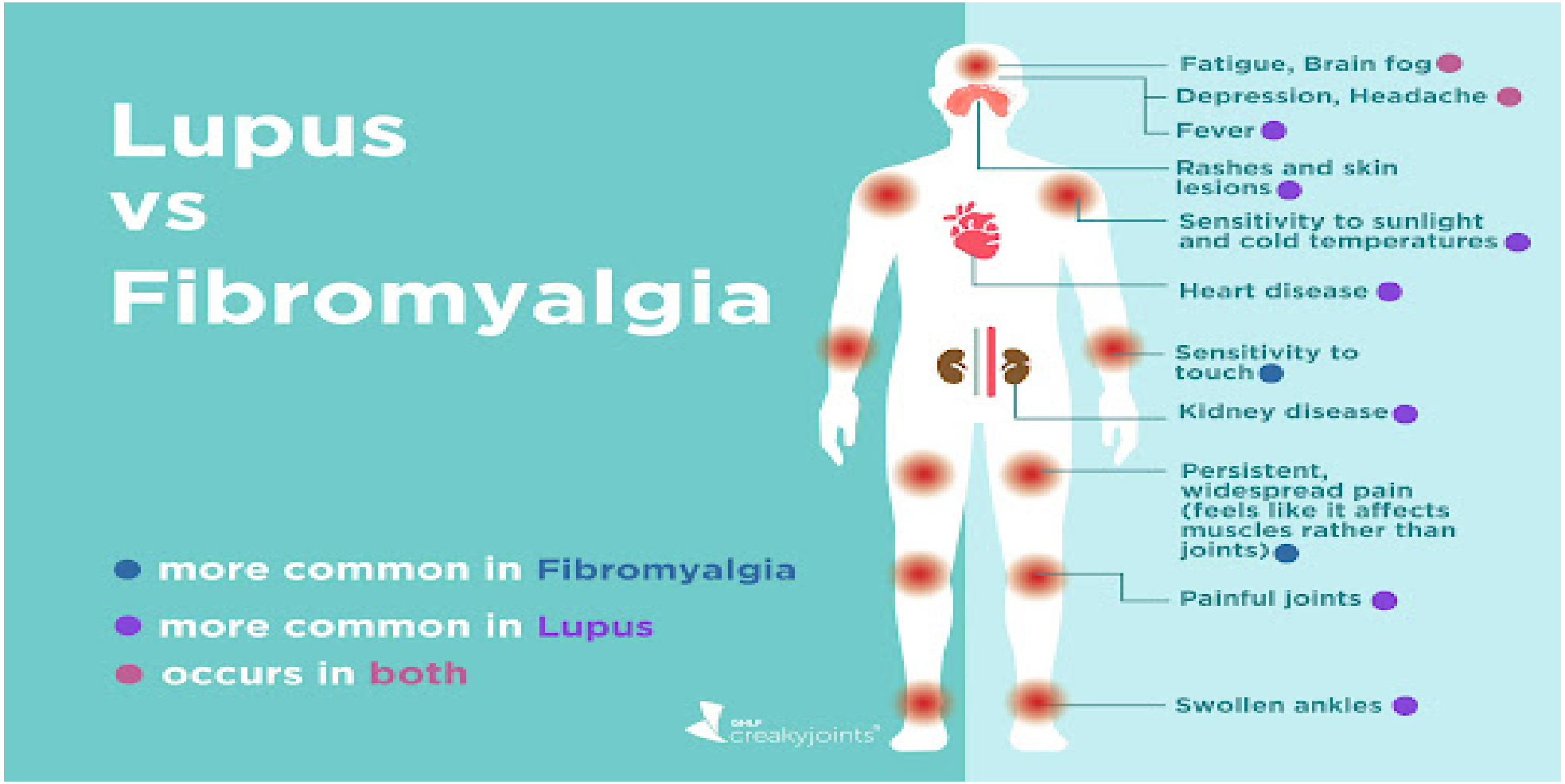
- Type I- Active SLE without FM- (SLEDAI >6, LN) – 30%
- Type II- Inactive SLE with predominate FM -8%
- Mixed SLE (active SLE with FM) – 13%
- Minimal SLE (Inactive SLE without FM)- 49%

* Rogers at al. March 2021

SLE types

- Type I- classic inflammatory features of SLE (arthritis, serositis, rash, nephritis)- respond to immunosuppression- measurable by SLEDAI, labs
- Type II- encompasses fatigue, impaired sleep, widespread pain, mood disorder and cognitive dysfunction- other tools to measure
- Implementing division of SLE in subtypes, encourages us doctors to address these important symptoms.

SLE types


Lupus
vs
Fibromyalgia
more common in Fibromyalgia
more common in Lupus
occurs in both
creakyjoints
Fatigue, Brain fog
Depression, Headache
Fever
Rashes and skin lesions
Sensitivity to sunlight and cold temperatures
Heart disease
Sensitivity to touch
Kidney disease
Persistent, widespread pain (feels like it affects muscles rather than joints)
Painful joints
Swollen ankles

Why is it important?

- Type II syndromes addressed in only about 30%
- Often perceived as SLE related symptoms
- Immunosuppression vs a more comprehensive approach, PT, sleep hygiene, pain management discussions

Questions? -
Remember- Rheumatologist is your Friend!