A Researcher's Guide to Lupus

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What is Systemic Lupus Erythematosus?

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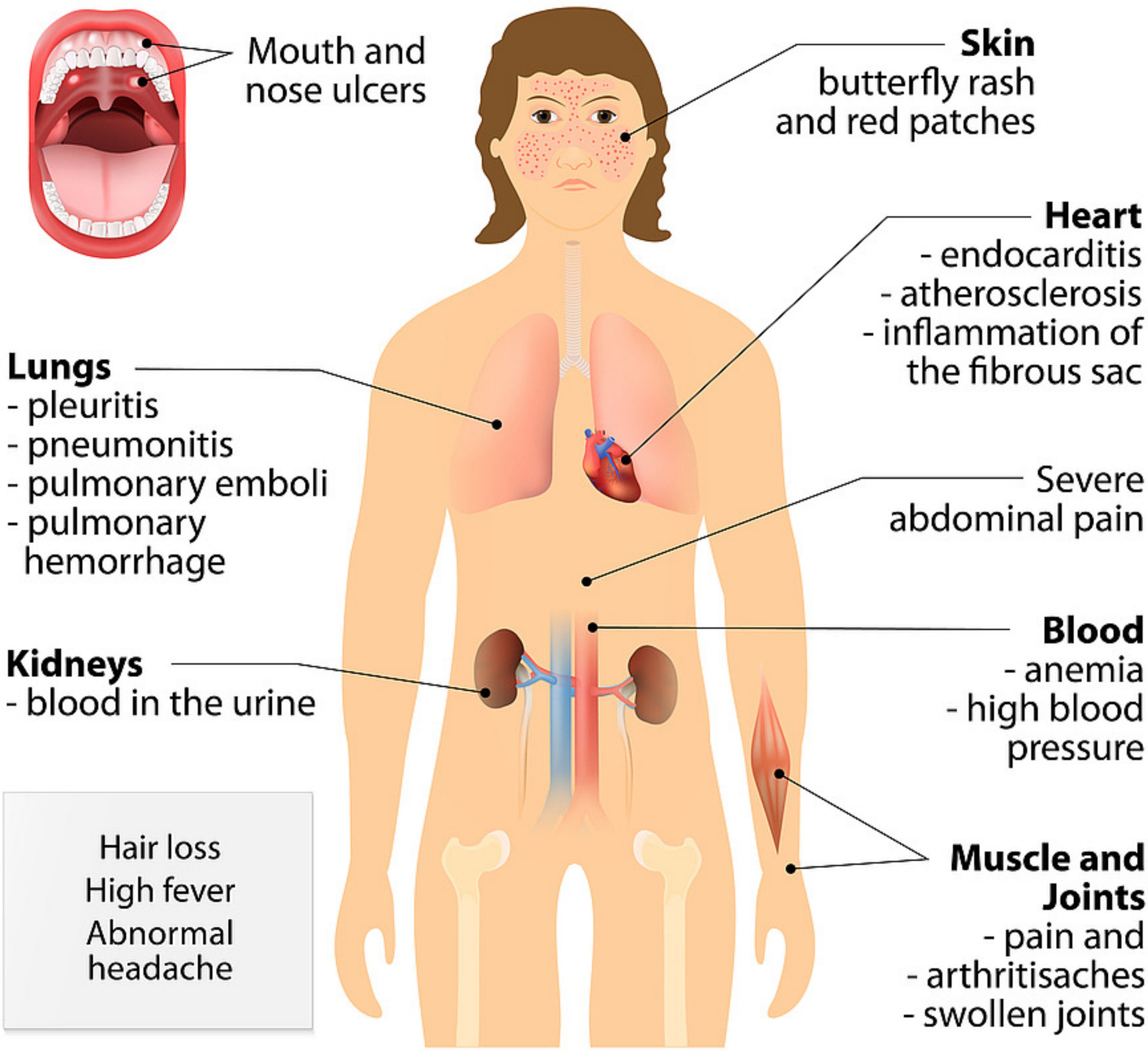

Systemic Lupus Erythematosus (SLE) is an autoimmune disease that is nine times more prevalent in women than men and two to three times more prevalent in African American, Hispanic, Asian and Native American populations than Caucasian. It is usually diagnosed in adults aged over 15 years old. It is very rare in children. In the United States, one estimate of the prevalence of SLE is 53 per 100,000; other estimates range from 322,000 to over 1 million.

Definitions

Active disease: Patient is symptomatic. He/she may or may not be experiencing a Flare- Up. Determined on-site by physician.

Inactive disease: "Periods of quiescence." Patient is still taking daily medication and does not have symptoms.

Flare-Ups: Patient is experiencing symptoms above their baseline symptoms. Typically, a lupus Flare involves all previous symptoms plus new ones at each Flare.

Mild: Symptoms are easily recognized and treated. i.e. – skin rash, headaches, fatigue, and mild pain.

Moderate: Abnormal blood counts – white or red blood cells. Typically this classification is blended with the severe category since blood involvement almost always indicates future organ system involvement.

Severe: Dx is affecting major organs/organ systems.

Actively Treated: Patient is receiving some form of treatment for Lupus-related symptoms. 94% of entire Lupus population is actively treated regardless of how active or inactive the disease is at the moment.

All comers: Both active and inactive Lupus patients.

Diagnosis

Required: 4 out of the following 11 criteria must be met:

(1) malar rash

(2) discoid rash

(3) serositis or pericarditis

(4) oral ulcers

(5) arthritis

(6) photosensitivity

(7) hemolytic anemia or leukopenia/lymphopenia or thrombocytopenia

(8) renal disorder

(9) antinuclear antibody (ANA) test positive

(10) immunologic disorder (positive anti-SM antibody, positive anti-dsDNA antibody and/or false positive syphilis)

(11) neurologic disorder

Diagnosis Additional Notes

Additional: Blood and organ function tests (blood cell count, renal function, liver function, neural function, musculoskeletal function) will be done to diagnose which form of SLE the patient currently has.

Special Note: SLE diagnosis is very tricky. Most patients live with SLE for 2 years before diagnosis. 90% of patients are diagnosed by a rheumatologist. SLE shares symptoms with many other autoimmune disorders (fibromyalgia, scleroderma, rheumatoid arthritis, chronic fatigue syndrome).

Primary and Secondary Symptoms

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Primary

- fatigue
- fever
- arthritic pain
- malar (“butterfly”) facial rash
- flaky spots on upper body
- sores/bumps on face & chest
- swollen glands
- light sensitivity
- headaches
- sudden chest pain
- hair loss
- swelling of the extremities

Secondary

- weight loss
- memory loss
- bloody urine
- numbness
- anemia
- anxiety/depression
- swelling of extremities
- complications in any organ

Patient Lifestyle

The disease can become active at any time, for any duration, and at any degree of severity. Typically, each Flare Up is worse (involves more symptoms) than the one before. No matter the degree of Activity vs. Inactivity, Lupus patients usually attend regular doctor visits increasing in frequency depending on the progress of their disease. A Lupus Status Test is NOT done at each visit, but rather only when the patient's health has altered. SLE patients usually keep their disease under control through medications and do not require laboratory blood work at doctor visits. Prognosis: there is no cure. Most patients have a 90% survival rate after more than 10 years. In 2013, around 70 % of patients survived.

Lupus Status Test

Blood: CBC (blood count, anemia indicates a flare); ESR (tests for inflammation, high ESR correlates with a flare); ANA is sometimes tested (presence of anti-DNAantibodies, specifically anti-dsDNA and anti-Smith protein, indicates a flare); C3 and C4 (low levels of these proteins indicate Lupus-related inflammation); creatinine and urea (increased levels indicate Lupus-related kidney damage); lipids (high levels indicate Lupus-related heart disease); liver enzymes (elevated levels indicate Lupus-related hepatitis or cirrhosis)

Urine: presence of blood cells or protein in urine indicate Lupus-related kidney damage. Urinalysis is rarely done for SLE patients

Bone: low bone density indicates Lupus-related osteoporosis. Bone density analysis is rarely done for lupus patients.

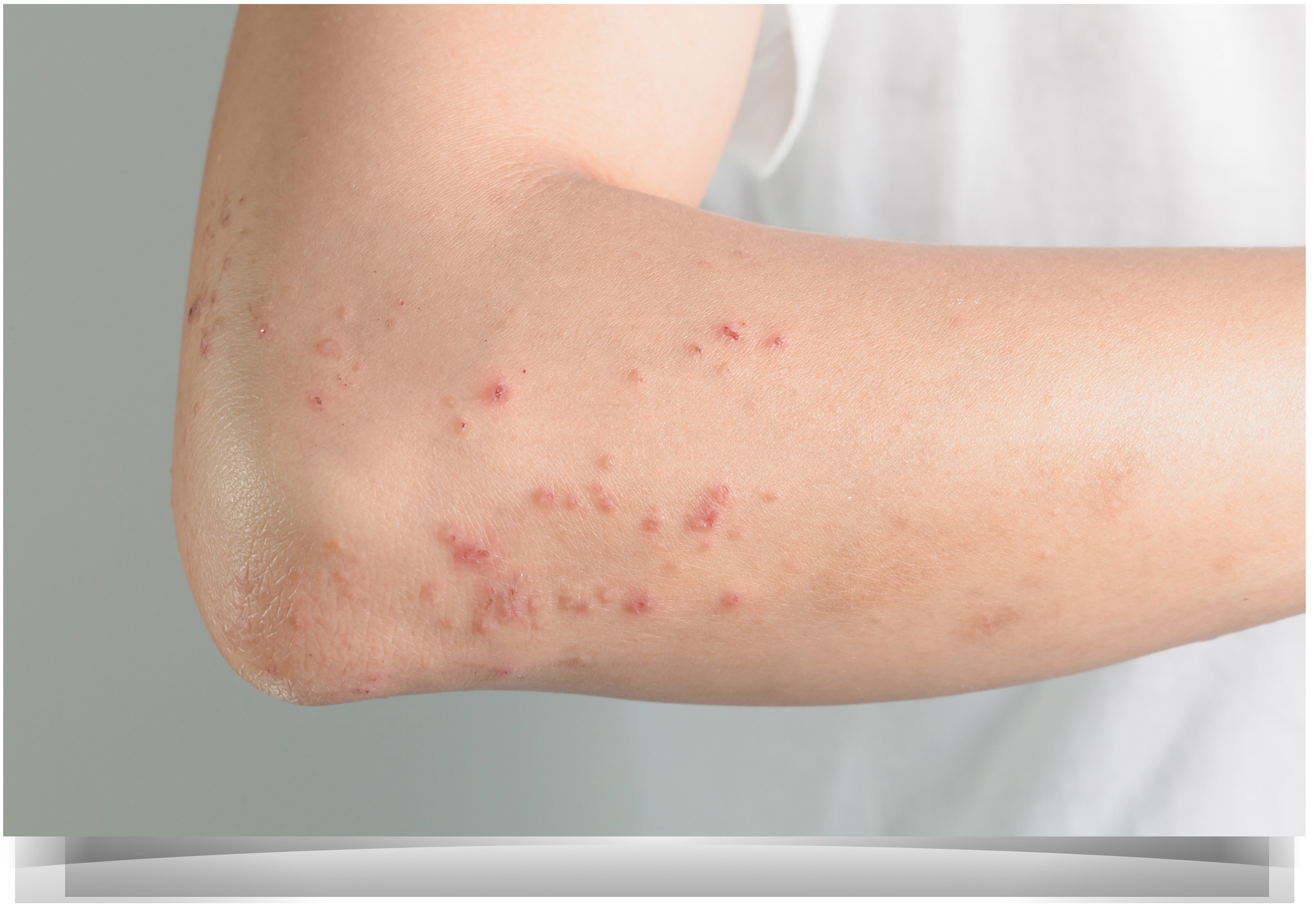
Flare Ups can be highly irregular and unpredictable. The average frequency is 3-6 per year and the average length can last an hour, several days or months, or even years.

Primary Treatment

The first treatment strategy is anti inflammation. The drugs most commonly used are NSAIDS. Specifically Ibuprofen (Motrin/Advil) naproxen (Aleve/Naprosyn) celecoxib (Celebrex). The dosage is usually low due to long-term Rx ”Low” varies depending on NSAIDs. Steroids, methyl-prednisolone (Medrol) and prednisone, are also used. The high dosage is for steroids is about 100-1,000mg, medium dosage is 15-100mg, and low dosage is 5-15 mg over a few days. Steroids are taken daily and doctors prefer to limit their prescription. Side effects may include: Bleeding ulcer, abnormal kidney function, stomach irritation blindness, muscle weakness, osteoporosis, high blood pressure, high glucose, and impaired blood clotting.

Secondary Treatment

The secondary stage of treatment is immuno-suppression. The medicines most commonly used are cyclophoshamide (Cytoxan), methotrexate (Rheumatrex), and azathioprine (Imuran). The standard dosage is 750mg/month over 6 months and high dosage is 50 mg/day over 4 days. The majority of patients maintain a standard regimen. The side effects are herpes zoster(Shingles), increased risk for cancer, increased risk for infection, hormonal inbalance, hair loss, sterility, bladder problems, liver cirrhosis, nausea, mouthsores, headaches, pancreatitis, and hepatitis.



Frequently Asked Questions

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What are the lines of treatment for SLE?

The traditional concept of "lines of treatment" does not directly apply to SLE due to the incredibly unpredictable and variable nature of the disease. However, anti-inflammatory drugs are the first and most often prescribed by rheumatologists. Second are the immunosuppressors.

What are the most common drugs taken for SLE?

Plaquenil is most common at our sites. Advil, Motrin, Celebrex, Aleve; prednisone; cyclophosphamide, methotrexate; Lovenox, Coumadin.

What is SLEDAI? What is BILAG?

SLEDAI, Systemic Lupus Erythematosus Disease Activity Index, is a scale designed to assess and categorize the level of disease activity in SLE. It is often used by researchers in clinical trials, but rarely, if ever, by the physicians treating lupus patients. It includes clinical tests (rashes, hair loss, etc.) and laboratory tests (blood tests, urinalysis, etc.) in the scoring index. Scores range from 0 to 105; a score greater than 20 is rare. On SLEDAI, a score of 6 or more corresponds to Active disease, though this is not what a physician will use to define Active disease. Physicians define Active as "symptomatic:" is the patient experiencing symptoms of their lupus or not? On SLEDAI, Mild/Moderate flare is defined as a change >3 points from patients previous score; Severe flare is a change >12.

BILAG, British Isles Lupus Assessment Group, is a score that attempts to asses a "need to treat" organ-by-organ. Scores range from A (very active) to E (not involved) and are broken into 8organ groups. As with SLEDAI, physicians do not use BILAG to assess their patient's level of disease activity.

What about longitudinal collections?

Since almost all SLE patients see their physician regularly, a longitudinal collection would be feasible

What are LE cells?

LE cells are immune cells that have "taken in" dead cells. The dead genetic material is visible as an LE body. In the past, LE cell presence was used to diagnose SLE, but it is no longer used as diagnostic criteria because only 50-75% SLE cases have LE cells.

What is cutaneous lupus?

Cutaneous lupus is an umbrella term for any type of lupus (SLE, discoid, drug-induced) that particularly manifests in the patient's skin. It can be SLE, discoid or drug-induced.

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Company Information

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