Help Us Solve
The Cruel Mystery

LUPUS™

FOUNDATION OF AMERICA

Living with Lupus

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Overview

- Knowing as much as you can about this complex disease
- Understanding how lupus can affect your body
- Making lifestyle changes to stay as healthy as possible
- Successfully managing lupus starts with awareness
  - Managing flares, changes in symptoms, physical conditions
  - Coping strategies

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Topics

- Lupus Clinical Features and Prognosis
- Treatment Considerations
- Lifestyle Considerations
- Coping Strategies
- Workplace and Resources
- LFA Resources

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What is Systemic Lupus Erythematosus?

Lupus is a chronic autoimmune disease that can damage any part of the body.

With autoimmune diseases, the body cannot tell the difference between “invaders” (i.e., bacteria and viruses) and the body’s own healthy tissues.

Lupus is…

- Different for each person
- A disease that ranges from mild to life threatening
- Not curable—yet , but symptoms of the disease may increase (flare), decrease, or become inactive (remission)

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Lupus is Not…

- Contagious or transmittable
- Like or related to cancer
- Like or related to HIV (Human Immune Deficiency Virus) or AIDS (Acquired Immune Deficiency Syndrome)

Who gets Lupus?

- **How common is Lupus in the US?**

  - More than 16,000 people in the U.S. develop lupus each year

  - Nearly 1.5 million people in the US

- **What ages are affected?**

  - Peak occurs between ages 15 – 40 y, but all ages are affected

- **Is everyone affected equally?**

  - 80-92% of US patients with lupus are women

  - Women of color are 2-3 times more likely to develop lupus

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Siegel M, Lee SL. *Semin Arthritis Rheum*. 1973;3:1–54 [Evidence Level C]; Rus V, Hochberg MC. In: Wallace DJ, Hahn BH, eds. *Dubois' Lupus Erythematosus.* 6th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2002. [Evidence Level C]

Lupus in the Hispanic & African American Communities

- **LU**pus in **MI**norities: **NA**ture versus Nurture (LUMINA) studies show that Hispanic and African American lupus patients tend to:
  - Have more severe disease overall
  - Develop lupus earlier in life
  - Experience greater disease activity at the time of diagnosis (including kidney problems)
  - Have more neurological problems such as seizures, hemorrhage (internal bleeding) and stroke.

Clinical Features

- Lupus is a chronic autoimmune disease that can affect virtually any part of the body.
- Lupus is much more common in women than in men. It can occur at any age.
- Lupus is a disease of spontaneous flares and remissions.
- The most common symptoms are fatigue, photosensitive rash, and joint pains.

American College of Rheumatology (ACR) Criteria for SLE

- Skin
  - Malar rash
  - Discoid rash
  - Photosensitive rash
  - Oral ulcers
- Arthritis
- Serositis
- Nephritis
- Neurologic disease
- **Hematologic disease**
  - Low white blood counts
  - Hemolytic anemia
  - Low platelets
- **Autoimmune antibodies**
  - Anti-double stranded DNA
  - Anti-Smith
  - Any antiphospholipid
- **ANA antibody**

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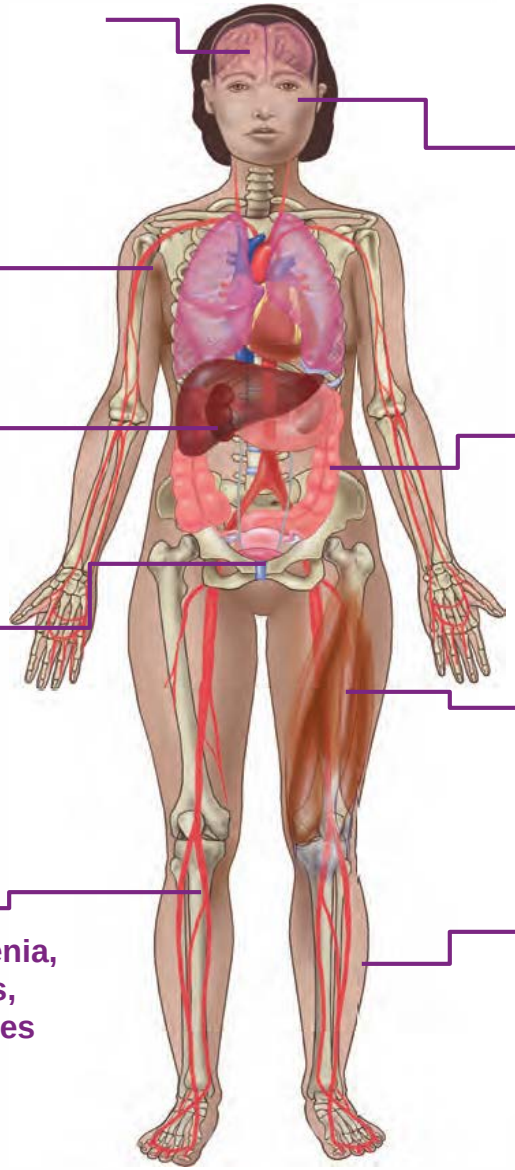
Hochberg MC. *Arthritis Rheum*. 1997;40:1725. [Evidence Level C]



Central and Peripheral Nervous System
Seizures, Psychosis, Headaches, Cognitive Dysfunction, Neuropathies, Depression, Low Grade Fever

Eyes and Mucous Membranes
Ulcers in the Eyes, Nose, Mouth or Vagina, Sjögren's Syndrome

Heart, Lungs
Pericarditis, Myocarditis, Endocarditis, Pleuritis, Pneumonitis

Kidneys
Edema, Hypertension, Proteinuria, Cell Casts, Renal Failure

Gastrointestinal
Nausea, Vomiting, Diarrhea, Weight Changes

Reproductive System
Pregnancy Complications, Miscarriages, Menstrual Cycle Irregularities

Musculoskeletal
Extreme Fatigue, Arthralgia, Myalgia, Arthritis, Myositis

Blood
Anemia, Thrombocytopenia, Leukopenia, Thrombosis, Circulating Autoantibodies and Immune Complexes

Skin
Butterfly Rash, Cutaneous Lesions, Photosensitivity, Alopecia, Vasculitis, Raynaud's Phenomenon

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How do we use Autoantibody tests?

- **Help diagnose SLE**
  - ANA antibodies: 99% SLE patients positive

§ **Direct attention to possible organ involvement**

—Anti-double stranded (ds)DNA: kidney disease

—Antiphospholipid: blood clots, miscarriages

—Anti-Ro (SS-A): rash, neonatal SLE

§ **Help to predict flares**

—Low complement C3, C4

—Anti-dsDNA

—Target labs (eg, Blood counts, urinalysis)

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McMahon M, Kalunian K. In: Lotze M, ed. *Measuring Immunity: Basic Science and Clinical Practice*. Oxford, UK: Elsevier Academic Press; 2005. [Evidence Level C]

Skin Manifestations of SLE

Malar rash (aka "Butterfly rash")

- 30%–60% of patients

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Klippel JH, ed. *Primer on Rheumatic Diseases.* 12th ed. Atlanta, Ga: Arthritis Foundation; 2001. [Evidence Level C]

Skin Manifestations of SLE

Discoid rash

- 15%–30% of SLE patients
- Occurs as part of SLE or in isolation
- (2%–10% of DLE patients develop SLE)

DLE = discoid lupus erythematosus.

Klippel JH, ed. *Primer on Rheumatic Diseases.* 12th ed. Atlanta, Ga: Arthritis Foundation; 2001. [Evidence Level C]

Skin Manifestations of SLE

Alopecia

- Diffuse or patchy
- Reversible or permanent scarring with discoid lesions
- “Lupus frizz”

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Klippel JH, ed. *Primer on Rheumatic Diseases.* 12th ed. Atlanta, Ga: Arthritis Foundation; 2001. [Evidence Level C]

Skin Manifestations of SLE

Photosensitivity

- Common in SLE patients: up to 58% of patients; 34% had flare of systemic symptoms
- Refers to flare of disease when patients are exposed to sun

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Wysenbeek AJ, et al. *Ann Rheum Dis*. 1989;49:461–463. [Evidence Level B]

Raynaud's Phenomenon

- Not a diagnostic criteria for SLE, but occurs in up to 60% of patients
- Triggered by cold or emotional stress
- Cold hands and feet associated with color change

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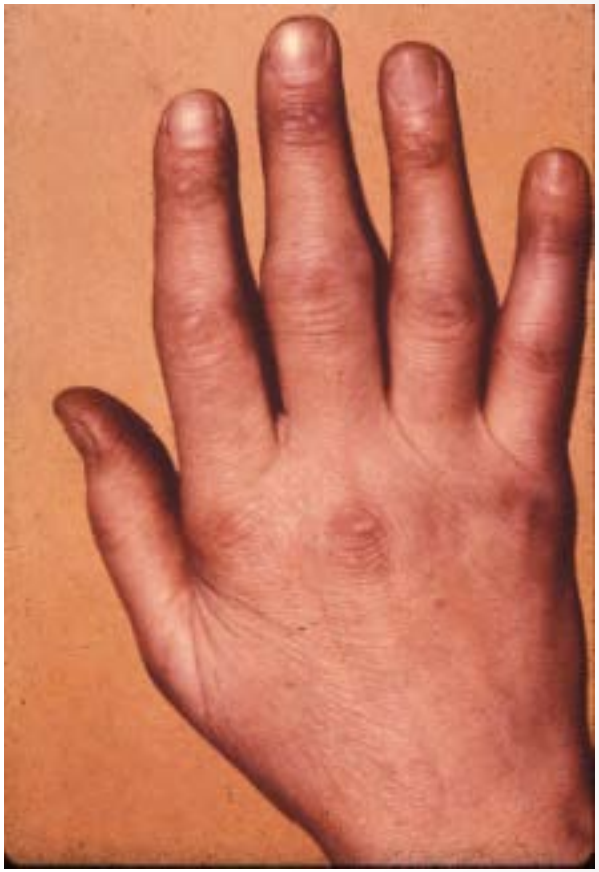
Klippel JH, ed. *Primer on Rheumatic Diseases.* 12th ed. Atlanta, Ga: Arthritis Foundation; 2001. [Evidence Level C]

Arthritis

- Between 76%–100% of patients
- Symmetric, usually involves small joints of hands, wrists, and knees, sparing spine
- Nondeforming, nonerosive



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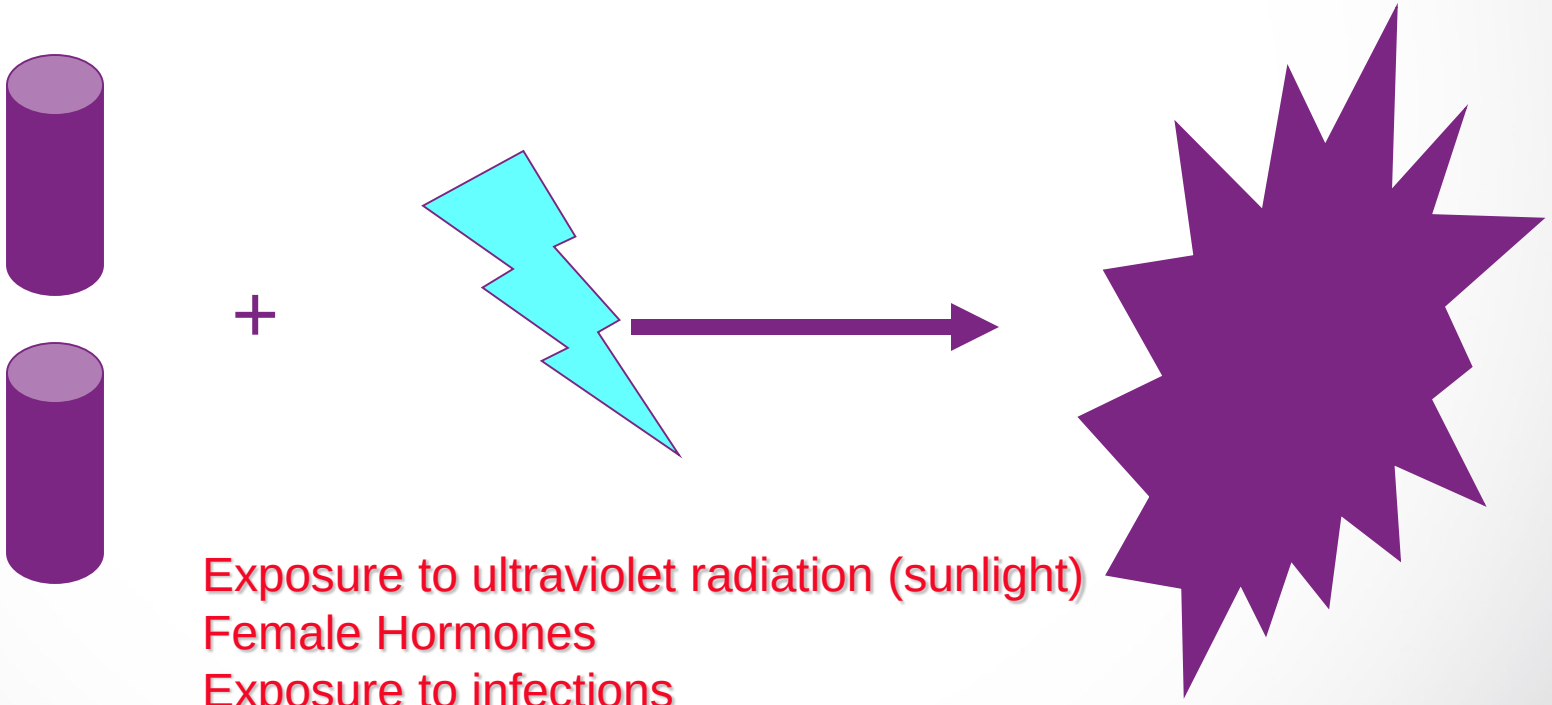
Klippel JH, ed. *Primer on Rheumatic Diseases.* 12th ed. Atlanta, Ga: Arthritis Foundation; 2001. [Evidence Level C]

What Causes Lupus?

GENES ENVIRONMENT ABNORMAL IMMUNE RESPONSE



Exposure to ultraviolet radiation (sunlight)
Female Hormones
Exposure to infections
Physical and emotional stress

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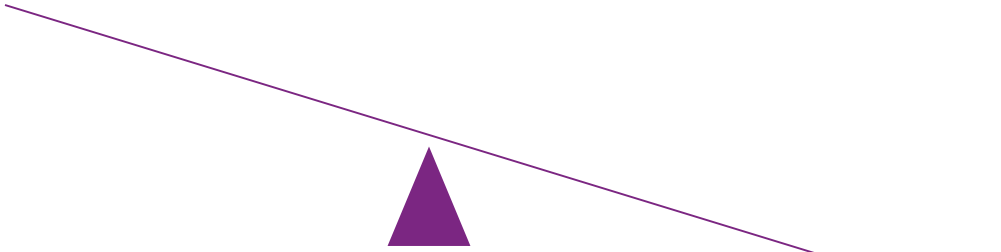
Immune Response



Immune Regulation

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Prognosis

- Most people with lupus can look forward to a normal lifespan:
  - Follow physician instructions
  - Know when to seek help for side effects or new symptoms
- Serious problems occur in some people and generally can be treated successfully
  - Close monitoring is essential
  - Take medications as prescribed

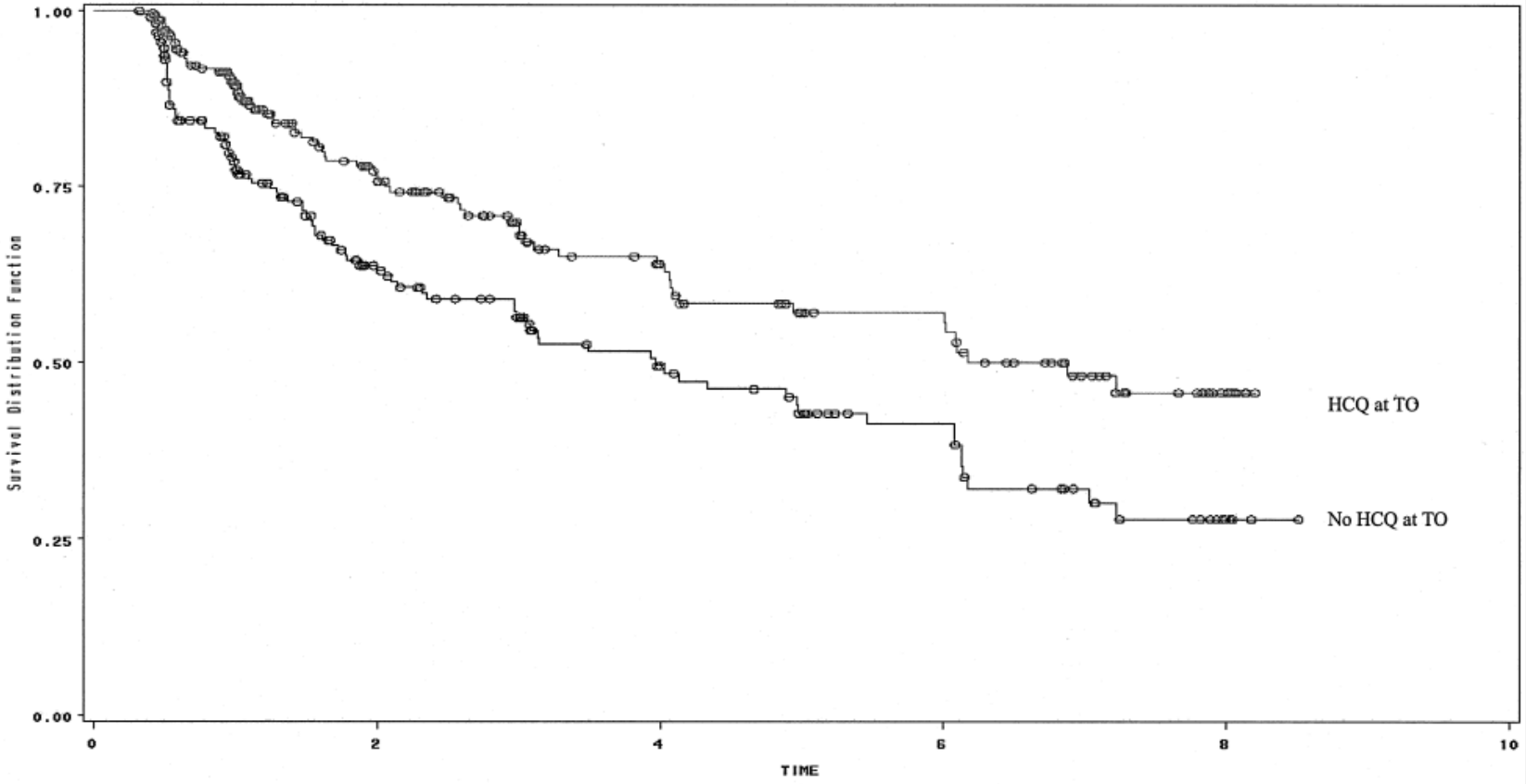
Treatment Considerations

- For most people proper treatment can
  - Reduce symptoms
  - Protect against serious organ damage
- Medications are an important part of managing lupus
  - Lupus is different for each person
  - Physician looks for the best combination of medications for you

Treatment of Mild-Moderate Lupus

- Rest
- Avoid sun exposure
- Topical corticosteroids
- Antimalarial drugs (plaquenil)
- Anti-inflammatory drugs
- Low-dose corticosteroids (prednisone)
- Health maintenance - preventative measures

Hydroxychloroquine Reduces Damage



Time (years)	0	2	4	6	8	10
HCQ at T0	297	105	57	41	9	0
No HCQ at T0	229	83	48	28	4	0

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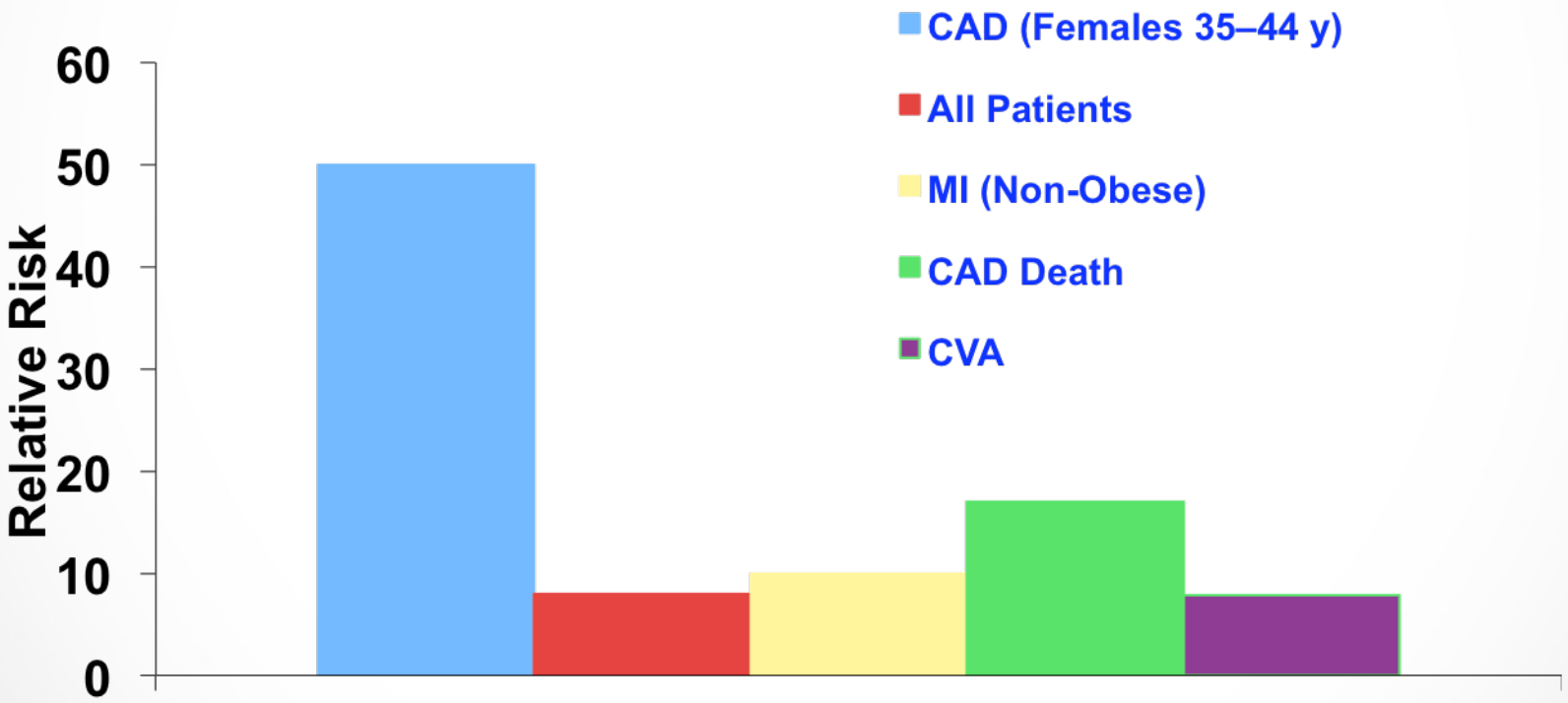
Fessler BJ et al, Arthritis Rheum, 2005

Treatment of Severe Lupus

- Often requires potent immunosuppressive drugs
- Close monitoring required for disease flares and/or medication side effects
- Consider participation in clinical trials

Long term complications of lupus: Atherosclerosis



•CVA = cerebrovascular accident.

Esdaile JM, et al. *Arthritis Rheum*. 2001;44:2331–2337 [Evidence Level B]; Karrar A, et al. *Semin Arthritis Rheum*. 2001;30:436–443 [Evidence Level C]; Manzi S, et al. *Am J Epidemiol*. 1997;145:408–415 [Evidence Level B]; Manzi S, et al. *Arthritis Rheum*. 1999;42:51–60. [Evidence Level B]

Lifestyle Considerations

- Fatigue
- Rest
- Exercise
- Diet and nutrition
- Smoking
- Sensitivity to light

Fatigue

- 80% of people with lupus experience significant fatigue
- It is unclear why fatigue occurs in so many people with lupus, but many factors appear to play a role
  - Disease activity
  - Pain
  - Medications
  - Poor physical conditioning
  - Mental health/Depression
  - Lack of social support

Rest

- Proper amounts of rest
  - Extremely important for people with lupus
  - At least 7 hours of sleep per night is a good goal
  - Helps with fatigue
  - Helps when experiencing flares
- Napping is helpful and sometimes necessary
  - Allow extra time in your day to rest

Diet and Nutrition

- There is no special diet for people with lupus
- A nutritious, well-balanced diet is advised
  - Fruits, vegetables, whole grains and moderate amounts of poultry and fish
- Avoid alfalfa sprouts, which may trigger flares
- Herbs, supplements and vitamins should be discussed with your physician

Diet and Nutrition

- Alcohol can be used in moderation, but may complicate the use of some medications
  - e.g., NSAIDs, anticoagulants, methotrexate
- Corticosteroids may elevate blood pressure and cholesterol – so fat and salt should be limited
- Due to the increased risk of osteoporosis, it may be helpful to eat foods rich in calcium
  - Green leafy vegetables, milk, cheese, yogurt, calcium supplements
- If fluid retention is a problem, salt should be limited

Smoking

- Tobacco is especially harmful to people with lupus
- Smoking impairs the body's entire circulatory system and increases the risk of atherosclerosis, a condition seen often in lupus patients
- Flares in cutaneous lupus have been linked to an ingredient in tobacco
  - Stop smoking and avoid second-hand smoke

Sensitivity to Light

- Two-thirds of people with lupus have increased sensitivity to ultraviolet (UV) rays
  - Sunlight, artificial light, or both

- Excessive exposure to the sun can cause a lupus flare
- Use sunscreen when outdoors
  - Both UVA and UVB protection
  - At least 50 SPF and reapply every few hours
- Consider sun protective clothing
  - For example: www.coolibar.com
  - www.sunprecautions.com

Sensitivity to Light

- Avoid sunlight between the hours of 10 am and 4 pm
- Clothing and hats also help protect from UV light
- For indoor light exposure, there are shields that cover fluorescent bulbs (can be ordered from several manufacturers)
  - Shields with nanometer readings of 380 to 400 are best

Coping Strategies

- Control fatigue:
  - Learn to pace activities and listen to your body
- Control stress:
  - Identify your stressors and use methods to control the stress
  - Say “no” when necessary
- Manage depression:
  - Most episodes subside on their own; inform your doctor if your depression is prolonged

Coping Strategies

- Manage anger:
  - Don’t let anger bottle up
  - Communicate with family/friends and help them understand what you are feeling
- Resolve guilt:
  - Modify thoughts and behavior
- Address intimacy issues:
  - Pain, fatigue, depression and certain medications can lower your interest in intimacy
  - Good communication helps counter negative self perceptions

What Can Lupus Patients Do to Decrease Their Risk of Heart Disease?

Ways to Improve ATH Risk Factors Without Medications

- Reduced intakes of saturated fat and cholesterol
- Exercise
- Weight control
- Stop smoking

Exercise

- Like everyone else, people with lupus should exercise regularly
- Exercise plans should be discussed with your physician
- Low impact activities are best if you're experiencing swollen joints or muscle pain
  - Walking
  - Swimming
  - Bicycling
  - Yoga/Pilates
  - Stretching

Exercise

- Regular exercise and even simple low-impact movement will:
  - Reduce or minimize stress
  - Help keep your heart healthy
  - Improve muscle strength and reduce muscle stiffness
  - Increase your range of motion
  - Help prevent osteoporosis

Exercise is good for your heart!

- The average number of carotid plaques is lower in SLE patients with high strenuous exercise levels and also with moderate and mild exercise levels
- Average IMT is also lower in SLE patients with strenuous and moderate exercise levels
- 58% of SLE patients who get low exercise had piHDL, compared to 44% of patients with medium to high exercise
  - Low exercise <225 mets/week
  - Medium exercise 225-945 mets/week
  - High exercise >945 mets / week

•*Volkmann et al. Arthritis Care and Research In press 2009*

Weekly Goals for exercise: Ideal >20 mets, minimum goal 12 mets

Exercise	METS per Hour	Exercise	METS per Hour
Light Pace		***Vigorous Pace***	
Walking slowly	2	Most doubles tennis	5
Walking average pace	3	Some exercise apparatuses	5
Light gardening	2	Some exercise apparatuses	5
Cooking, Light housekeeping, shopping	2.5	Slow jogging (one mi every 13 to 14 min)	5
Golfing with cart	3	Ice or roller skating	6
Bowling	3	Hiking	6-7
Moderately Vigorous Pace		Bicycling 10 to 16 mph	6-10
Weight lifting, water aerobics	3.5	Swimming laps moderately fast to fast	6-10
Cycling (<10 miles/hr)	4	Jogging (1 mile every 12 min)	8
Walking at a brisk pace (1 mi every 20 min)	4	Running 8 mph (7.5-minute mile)	13.5
Dancing	4	Running 10 mph (6-minute mile)	16

http://healthfullife.umdnj.edu/archives/exercise_health_archive.htm

Approaching the concept of "Treat to Target" in SLE

LLDAS: A state of SLE disease activity that, if sustained, leads to lower likelihood of adverse outcomes:

1. SLEDAI ≤ 4 without major organ activity
2. No new disease activity
3. PGA (0-3) ≤ 1
4. Prednisone equivalent dose ≤ 7.5 mg/day
5. Stable dose of maintenance treatments

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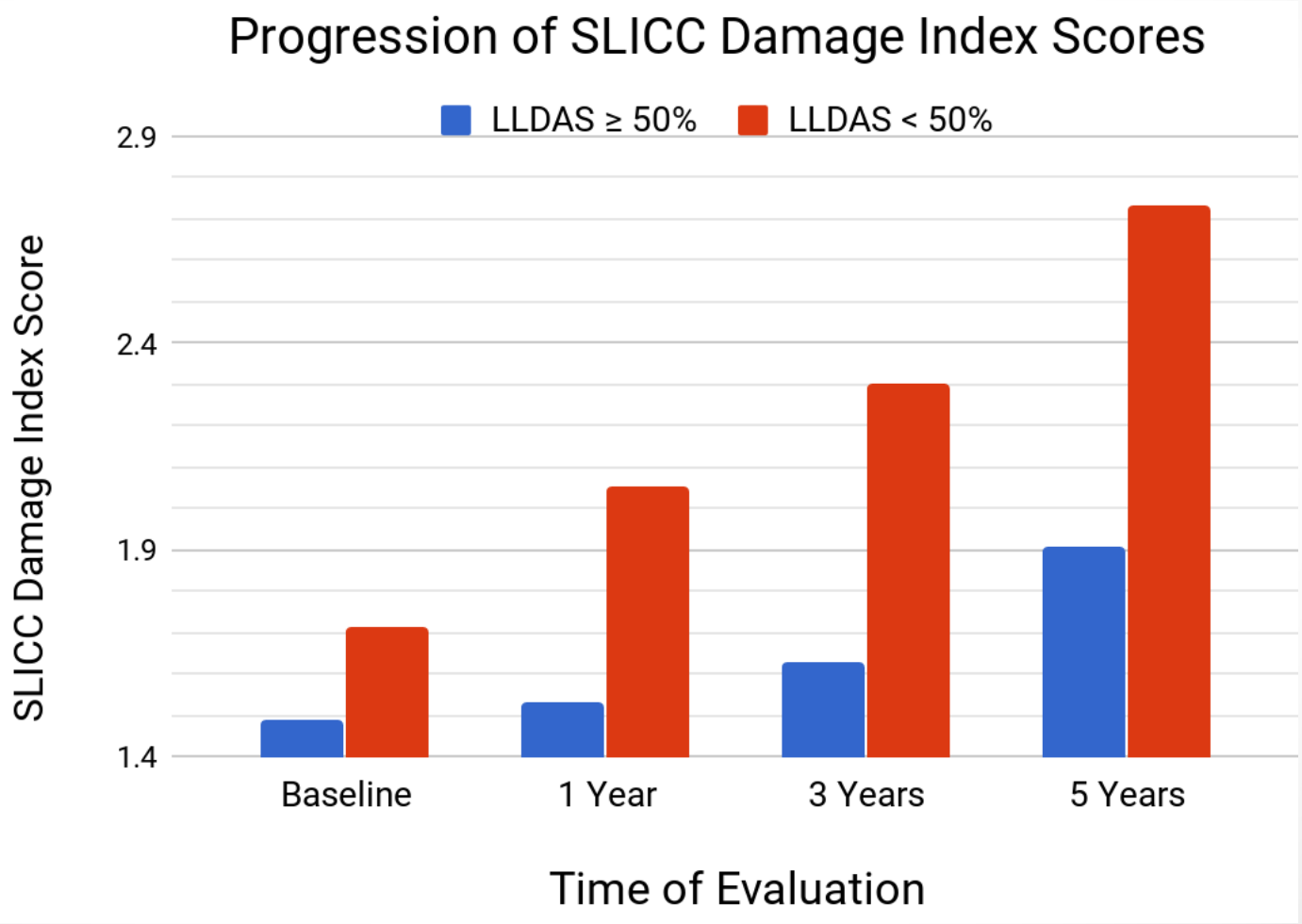
(Franklyn, K., et al. *Ann Rheum Dis*. 2016; 75: 1615-1621)

Damage Progression in SLE is lower when patients are in LLDAS >50% of the time



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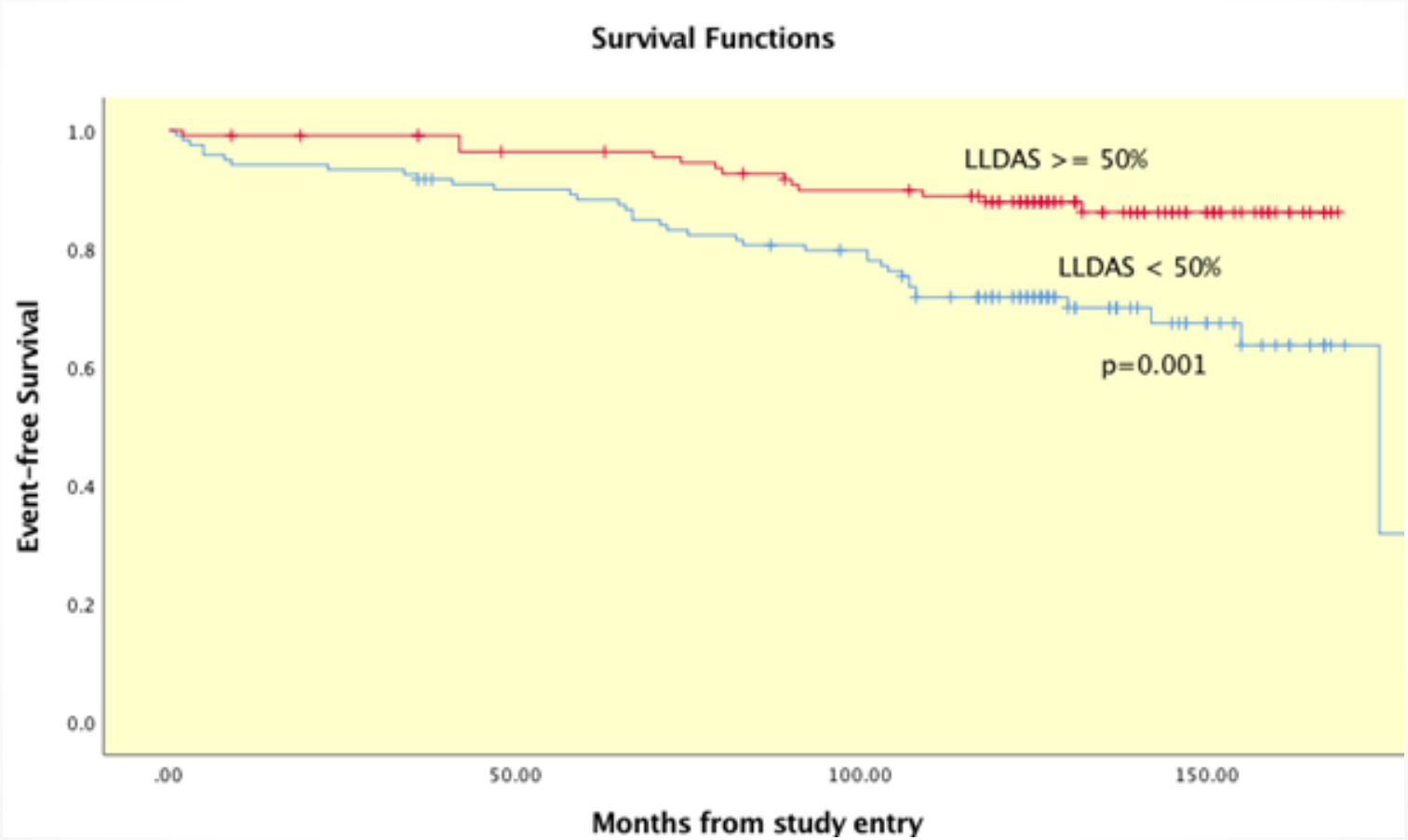

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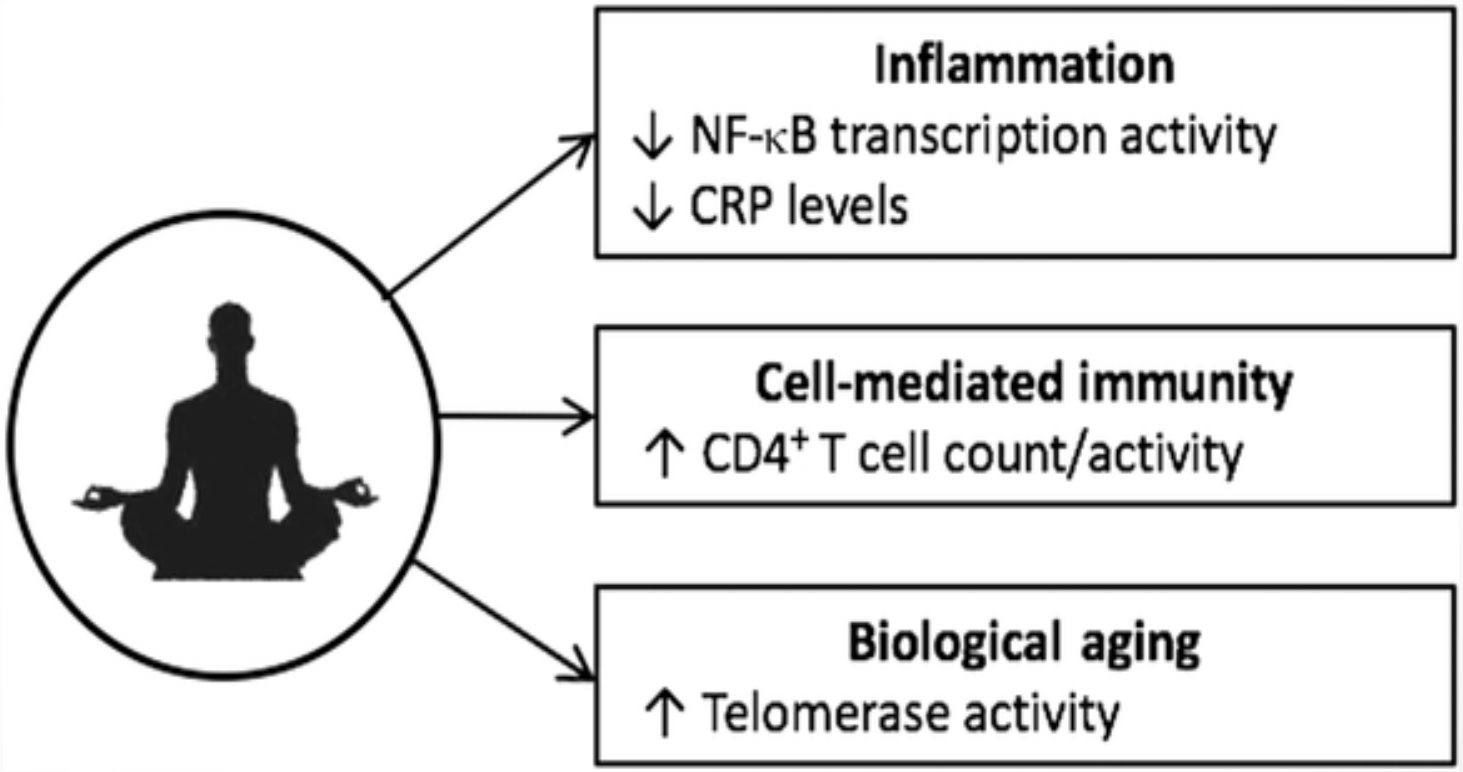
Yoga can improve your health and well being

- Yoga has been shown to improve pain, stiffness, swelling and physical and psychological health in patients with arthritis
- Also moderate evidence that yoga helps fatigue and symptoms of fibromyalgia

•Cheung et al Am J Phys Med Rehab 2016; Moonaz J Rheum 2015

Mindfulness meditation and the immune system: a systematic review of randomized controlled trials



•**Annals of the New York Academy of Sciences**
21 JAN 2016 DOI: 10.1111/nyas.12998
http://onlinelibrary.wiley.com/doi/10.1111/nyas.12998/full#nyas12998-fig-0001

Remember…

- There is more to you as an individual than having lupus
- Allow for personal time
- Adhere to medication regimes
- Communicate
  - It’s okay to say no
  - Be open with loved ones and with your physician(s) and other health care providers

Also Remember….

- Continue to learn as much as you can
- Incorporate lifestyle changes
- Manage lupus with awareness
  - Symptoms
  - Flares
  - Tension and stress
- Use coping strategies identified here

On the Horizon

- There has been enormous progress in recent years in understanding the biology of lupus.
- Numerous new approaches to treatment are under development and/or investigation.
- Information on participating in clinical trials can be found on the LFA web site at: www.lupus.org/searchclinicaltrials

Current Trials and Studies in SLE:

Anifrolumab- anti-Interferon alpha- TULIP studies
-in general lupus and lupus nephritis

Baricitinib in Lupus (UCLA site)

Ustekinemab in Lupus

Current Trials and Studies in SLE:

Belimumab in Early Lupus (UCLA site)

ALE-09: cannabinoid derivative in lupus

PREDICTS: observational study of markers of Heart Disease in Lupus

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Lupus Foundation of America Resources

- Visit the LFA at **www.lupus.org**
  - LFA Network of Health Educators available to answer questions by email or phone
  - Online resources and print materials
  - LupusConnect – online lupus community
  - Find a local support group
    - Support groups can also help you find local community health resources
  - Ways to get involved (Walks, Advocacy, Facebook, Twitter, etc.)

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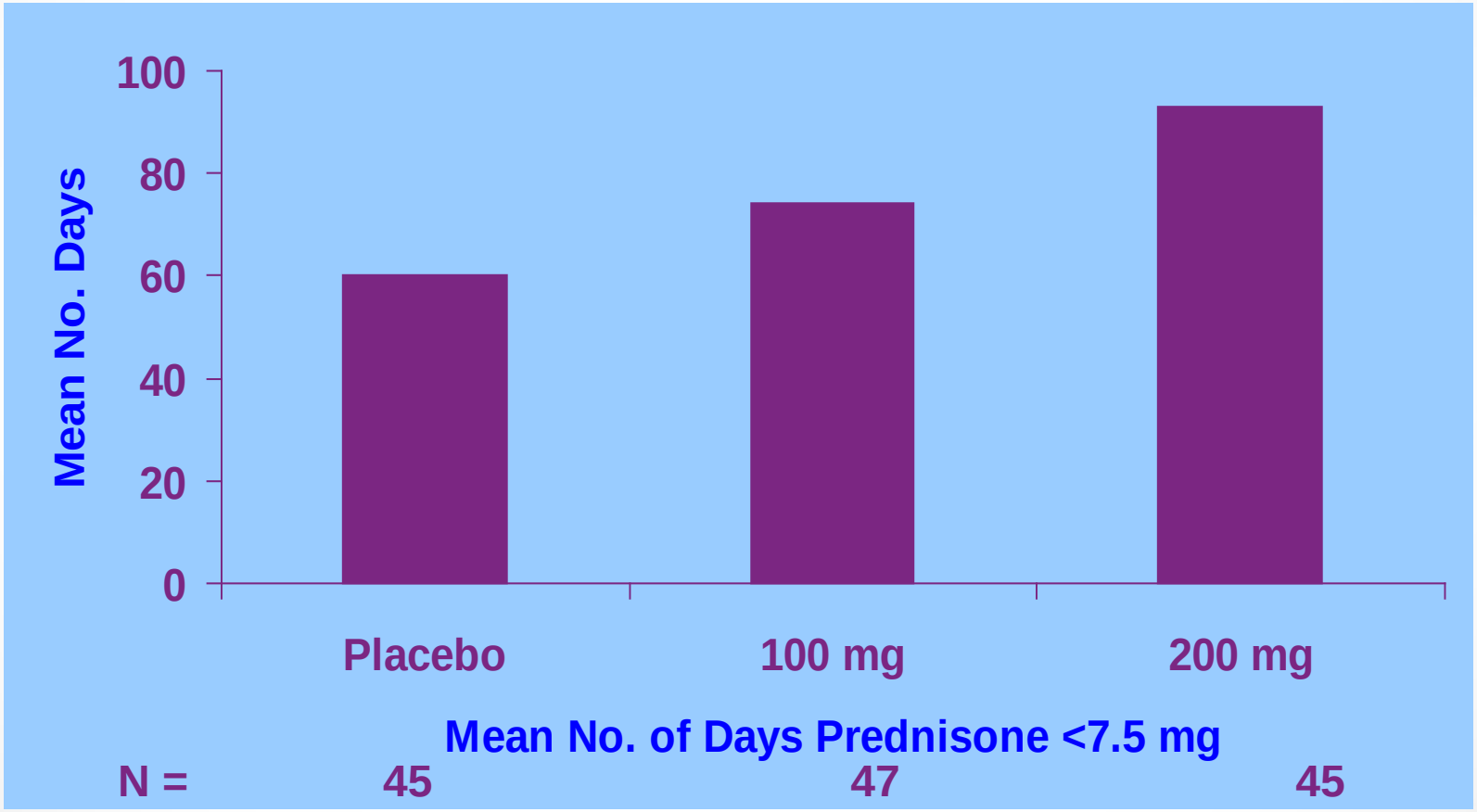
DHEA 200 mg Reduces Prednisone Requirement



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Petri MA, et al. *Arthritis Rheum*. 2002;46:1820–1829. [Evidence Level A]

DHEA in SLE

- A recent study also suggested that DHEA may help protect against bone loss in women who are also taking prednisone
- However, a recent randomized clinical trial found DHEA resulted in no improvement in Fatigue

Hartkamp, AnnRheum Diseases 2010; Sanchez-Guerrero J Rheum 2008

Belimumab: First FDA approved drug for SLE since 1955

- Approved for use in Adult patients with Active SLE who are currently receiving standard therapies
- Not approved for use in severe active lupus nephritis or severe CNS lupus
- Not approved with other biologics or iv cyclophosphamide

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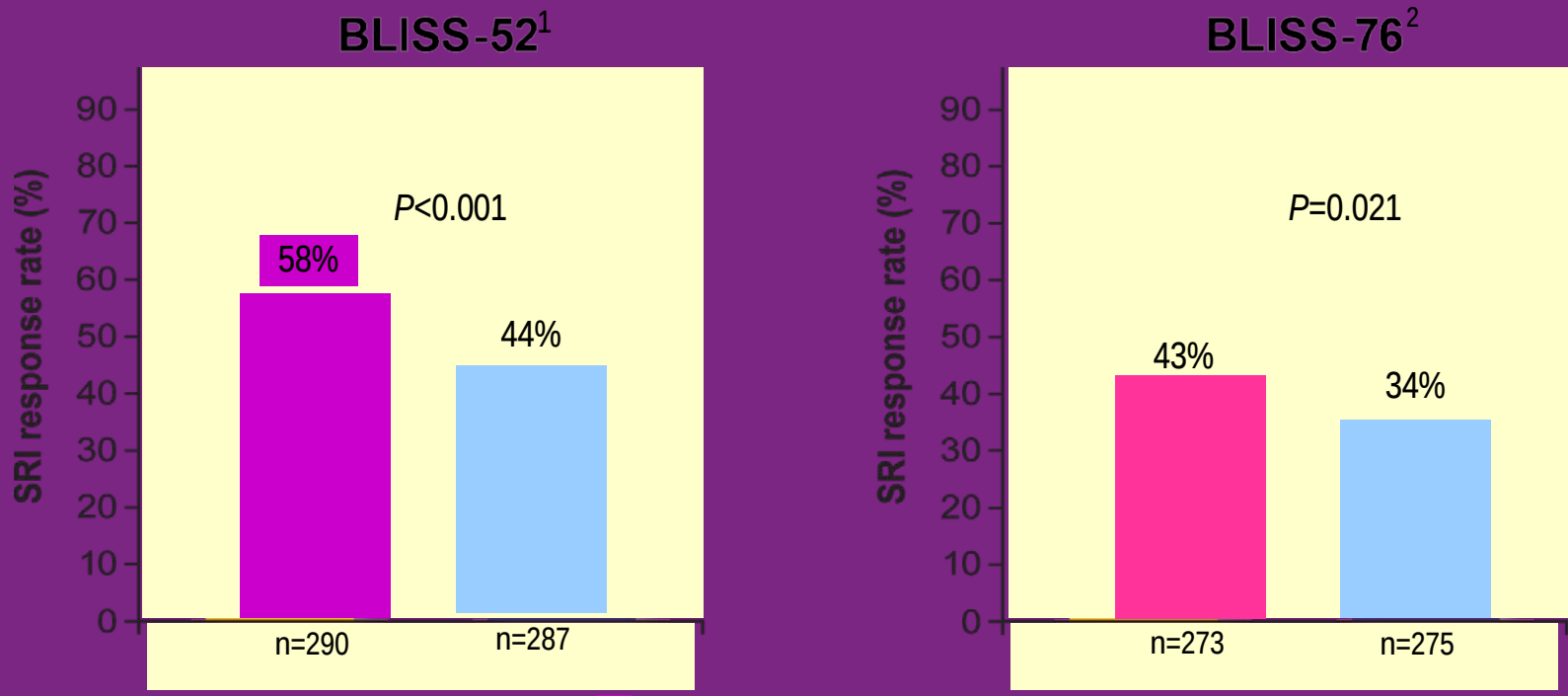
http://www.fda.gov

Belimumab in SLE: Percentage of Patients Meeting SRI Response Criteria at Week 52



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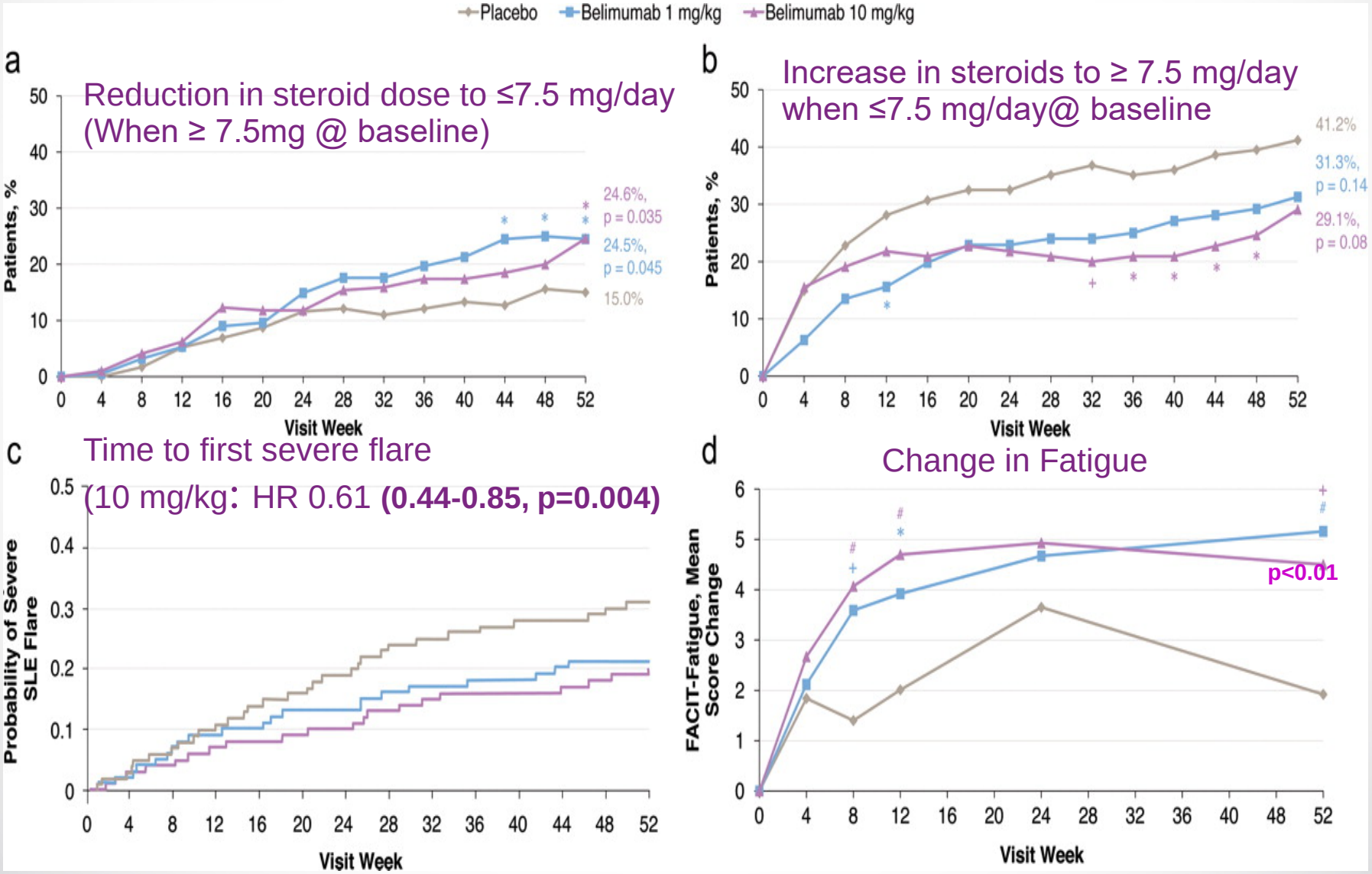
1. Navarra SV, et al. *Lancet.* 2011;377:721-731. 2. Furie R, et al. *Arthritis Rheum.* 2011;63:3918-3930.

BLISS trial secondary endpoints in pooled low complement/anti-dsDNA-positive subgroup.



van Vollenhoven R F et al. Ann Rheum Dis 2012;71:1343-1349