

Systemic Lupus Erythematosus: Living With Lupus

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A Comprehensive Guide for Patients and Families

Topics

- What is lupus?
- What causes it?
- How is it treated?
- What can I do to stay well?

What is SLE?

- An autoimmune disease
 - Auto = self
 - Immune system = the part of our body that protects us from invading germs, cancer, etc.
- Can attack any part of the body

SLE: meaning of words

- **Systemic**: it can affect any system of the body (joints, heart, lungs, nerves, kidneys, etc.)
- **Lupus**: Latin for wolf
 - Rash on face is like a wolf scratched and bit the face
- **Erythematosus**: medical term for red
 - Most lupus rashes are reddish in color

malade si lamenerent droit a leglise saint martin ou il voulut faire
la nonaine. Quāt il y fut si fist ses oraisons Et les chanoyues alle-
rēt faire le seruice du glorieux saint Et depuis fut leuesq en leglise en
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Commet saint martin accompaignie de saint brix sapparut
a leuesque du liege en son dormāt ⁊ le guerit de la maladie



Et la nuyt sicomme il sommeilloit vit mon seigneur saint marti
reneustu de moult beaulx et nobles vestemens pontificaulx et
sembloit bien arceuesque et decouste lui estoit saint brix cōme euesque
qui lui tenoit compaignie. Et lors saint martin dist a saint brix
Cest euesque du liege cy p̄sent est venu en mon eglise de moult loig
fain pays moy requerir guairison Si est bien droit quil lait car il la
bien desseruy Saint brix lui respondist que si fesoit son plaisir quil
feroit biē ⁊ quil estoit homme de bonne vie Et tātost mō seigneur saint

St. Martin
cured
Bishop of
Liege of
lupus
10th Century

Lupus: Types

- Cutaneous lupus
 - Only affects the skin
- Neonatal lupus
 - Affects newborn babies
- Drug-induced lupus
 - Resolves when offending drug is stopped
- Systemic lupus erythematosus (SLE)
 - Can affect any part of the body

What causes SLE?

- People are born with the genes which cause it.
 - Identical twin = 25% chance of having SLE if 1 has it.
- Something in the environment must “turn on” these genes
 - Ultraviolet light (sun)
 - Low vitamin D levels
 - Smoking
 - Viral infections (mono, Epstein Barr, parvovirus, etc)
- Hormones
 - 90% of SLE patients are women

SLE: how it is diagnosed...

need 4 out of 17 criteria

- Acute or subacute cutaneous lupus
 - E.g. butterfly rash (malar rash)
- Chronic cutaneous lupus (e.g. discoid lupus)
- Oral or nasal ulcers
- Nonscarring alopecia (hair loss)
- Inflammatory arthritis
- Serositis
 - Pleurisy
 - Pericarditis
 - Peritonitis

SLE: how it is diagnosed...

need 4 out of 17 criteria

- Kidney involvement
 - Protein in urine
 - Or red blood cell casts in urine
- Nerve involvement
 - Seizures, psychosis, neuropathies, spinal cord inflammation, brain inflammation
- Hemolytic anemia
- Low white blood cell counts
- Low platelet counts

SLE: how it is diagnosed...

need 4 out of 17 criteria

- ANA
- Anti-ds DNA
- Anti-Smith
- Antiphospholipid antibodies
- Low complement levels
- Positive Coombs antibody

Discoid lupus rash



(a)



(b)

Discoid lupus hair loss



Butterfly (malar rash)



Butterfly (malar rash)



Rash due to the sun: photosensitive or sun-sensitive rash



How is SLE treated?

- Avoid substances which make lupus worse
 - Protecting skin from ultraviolet (UV) light
 - Not smoking cigarettes
- Use medications to “calm down” the immune system

How is SLE treated?

■ Protecting self against UV light

- Daily sunscreen use
 - SPF 50 or better
 - Protects against UVA and UVB rays
 - Reapply 2-3 times on sunny days
 - Use Rit's Sun Guard when wash clothes
- Avoid sun between 10AM – 5PM
- Wear wide brimmed hat

How is lupus treated?

- Avoid substances that make lupus worse
 - Sulfa antibiotics
 - Septra, Bactrim, Trimethoprim-sulfamethoxazole
 - Always list sulfa antibiotics as an allergy!
 - Echinacea
 - Herbal product used to treat colds
 - Alfalfa/mung bean sprouts
 - Amino acid L-Canavanine can cause lupus flares

How is lupus treated?

- Medicines that “calm down” the immune system
 - **Anti-malarial medicines**
 - All SLE patients should be on one
 - Plaquenil (hydroxychloroquine)
 - **Steroids**
 - Prednisone and Medrol = most common
 - Fastest working medicines
 - Life saving
 - **Immunosuppressant medicines**

Anti-malarial Medications

- Plaquenil (hydroxychloroquine)
- Aralen (chloroquine)
- Atabrine (quinacrine)

Anti-malarial Medications

- WWII sailors took medicine to prevent malaria
- A Navy doctor noted that those with lupus and rheumatoid arthritis got better
- “Calm down” the immune system
 - Without suppressing the immune system

Anti-malarial Medications

- Especially good for:
 - Arthritis, rash, pleurisy, fatigue
 - Low blood cell counts
 - **Decreasing the need for prednisone**
 - **Decrease the risk of severe organ damage**
 - **Decrease death**
 - **Decrease blood clots**
 - **Improve pregnancy outcomes**

How is SLE treated?

- Plaquenil

- All SLE patients should take Plaquenil
- Very rare to have any major side effects

Plaquenil :

Side Effects

- Common (10%)
 - Rash
 - Headache
 - Dizziness
 - Insomnia
 - Stomach upset
- Usually resolve with a lower dose
- Can start at a lower dose and go up slow to make it tolerable

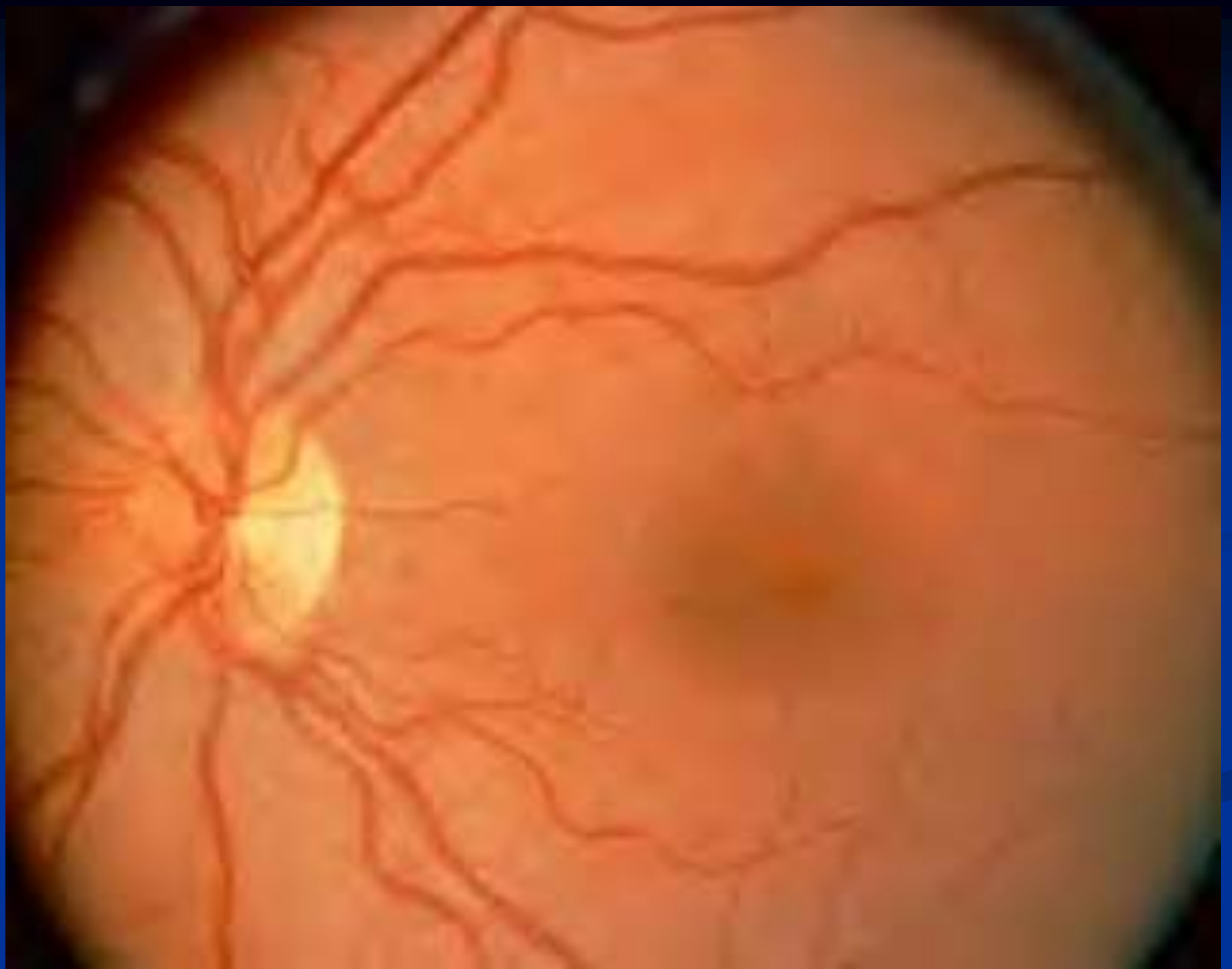
Plaquenil and Chloroquine: Eye damage!

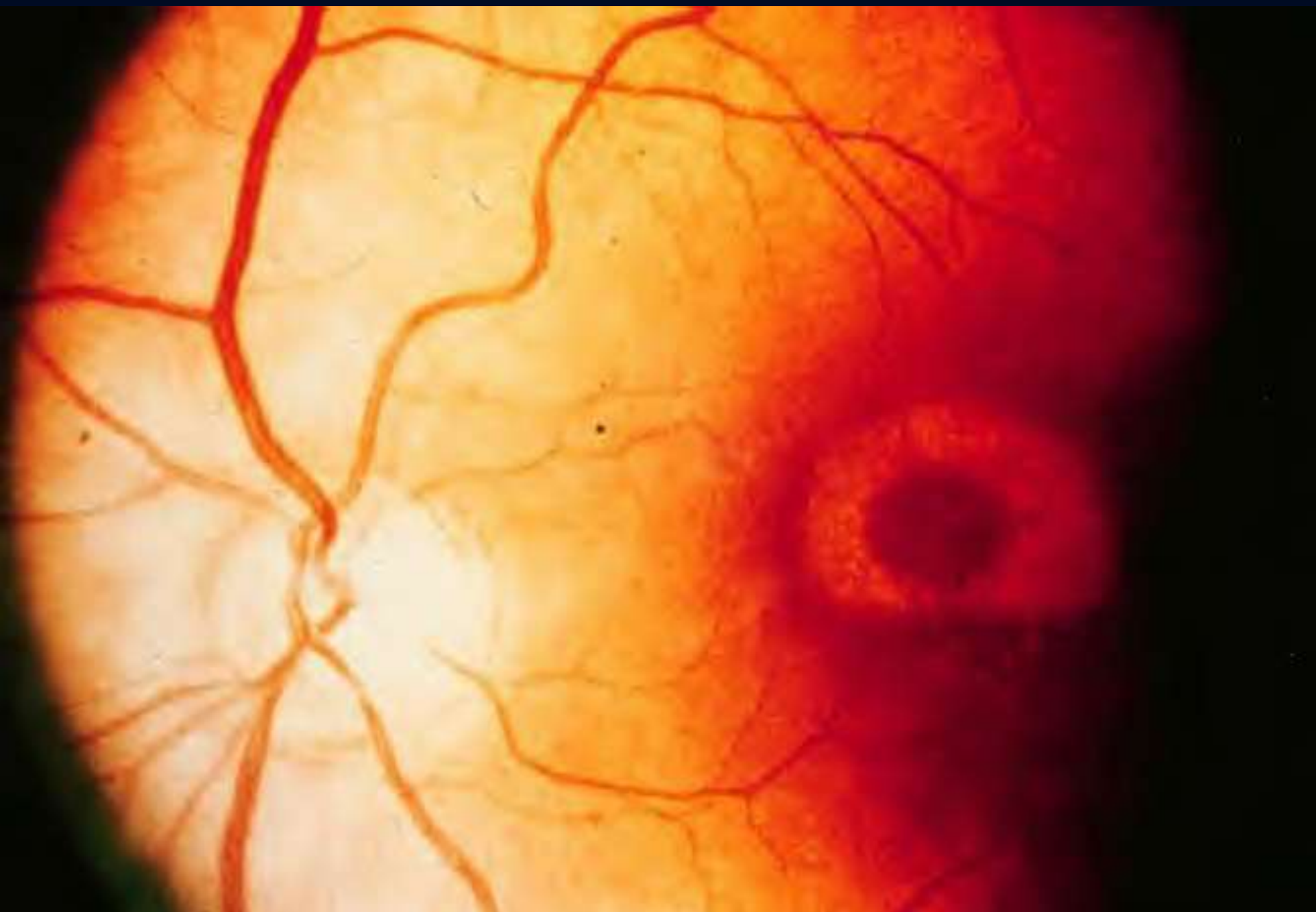
- The medicine can deposit on the back of the eye (retina)
- Causing vision loss
- Uncommon with Plaquenil
- More common with Chloroquine
 - 10% of users

How to prevent eye damage:

■ **Plaquenil and Chloroquine**

- See an ophthalmologist regularly
 - Yearly for Plaquenil
 - Every 3-6 months on chloroquine
- Insist that you get 2 of the following tests yearly
 - VF 10-2 (visual field test)
 - SD-OCT
 - mfERG
 - FAF





Smoking and Plaquenil

- Smoking cigarettes decreases the effectiveness of Plaquenil.



Steroids

- Naturally occurring hormones
- Most commonly used:
 - **Prednisone**
 - **Medrol** (methylprednisolone)
- Decrease inflammation quickly
- Suppress immune system at high doses
- Body normally makes 5-7 mg prednisone daily (4-6 mg Medrol)

Steroids

- Discovered in 1949
- Life saving medication
 - Before steroids, most lupus patients died within 2 years
 - Before steroids, almost all children with lupus died during the first year after the diagnosis

Prednisone: Side Effects

- Occur in most people
- Less likely to occur with prednisone 6 mg a day or less
- 20mg a day or more considered high dose
 - Increased risk for infection and other side effects
- Less side effects if taken 1st thing in the AM

Prednisone: Side Effects

- Increased appetite
- Weight gain
 - Breaks down muscle
 - Increases fat
 - Fluid retention
 - “moon face”
 - “buffalo hump”
- Skin thinning and bruising
- Increased hair growth or hair loss







Prednisone: Side Effects

- High blood pressure
- Heart disease
- Diabetes
- Osteoporosis (soft bones)
- Cataracts and glaucoma
- Depression and anxiety
- Infections

Prednisone: Side Effects

■ Adrenal insufficiency

■ Adrenal gland

- a gland on top of the kidney
- It makes prednisone-like steroids
- These are very important for living
- Makes more steroids during times of stress and infection
- Regular prednisone use causes the gland to stop making its own steroids

Prednisone: Side Effects

- Adrenal insufficiency
 - Need to take extra prednisone (up to 20mg a day) during times of stress:
 - Surgery
 - Infection
 - Trauma
 - Consider wearing a medical alert bracelet

Prednisone:

Tips on avoiding side effects

- Never miss a daily dose
- Always take it first thing in the morning
 - If you miss a dose take it later in the day
- Must be tapered down slowly
- Always work with your doctor on trying to get on the lowest dose possible
- Wear a medical alert bracelet

Prednisone:

Tips on avoiding side effects

- Weight gain

- Exercise regularly

- Toning exercises 3 days a week
 - Aerobic exercise 4-5 days a week

- Strict diet!

- Low calorie
 - Low fat
 - Low cholesterol
 - Low salt (sodium)

Prednisone: avoiding side effects

■ Preventing osteoporosis

- Need 1200 – 1500 mg calcium a day
 - Glass of skim milk = 300 mg
 - 8 oz yogurt = 300 mg
 - 2 cups cottage cheese = 300 mg
 - Calcium supplements
- Need 400-800 IU Vitamin D a day
 - Vitamin D milk
 - Calcium with vitamin D supplements
 - 1-2 multiple vitamins a day

Prednisone: avoiding side effects

- Preventing osteoporosis
 - Weight bearing exercise 30 minutes 5 days a week
 - Walking
 - Treadmill
 - Aerobics
 - Weight training
 - If on 5 mg a day or more for more than a few weeks:
 - Take Fosamax, Actonel, Reclast, or Boniva
 - Get a regular bone density x-ray yearly to monitor

Immune suppressant medicines

- Prednisone (over 7 mg a day)
- Methotrexate
- Arava (leflunomide)
- Imuran (azathioprine)
- Cytoxan (cyclophosphamide)
- CellCept (mycophenolate)
- Biologics
 - Benlysta, Rituximab, Remicade, Enbrel, Humira

Immune suppressant medicines: side effects

■ Infection

- Always call and see your primary care physician ASAP if you develop fever, cough, sore throat, painful urination, etc.

■ Lowered blood counts (except prednisone)

- Need a blood test (CBC) every 4-12 weeks

■ Liver problems (except prednisone)

- Need a blood test (LFTs) every 4-12 weeks

Important things to remember when taking immunosuppressants:

- See doctor at 1st symptoms of infection
- Medicine should be stopped during infection
 - Except prednisone
- Should be stopped before and right after surgery
 - Except prednisone, which should be continued
- Blood tests need to be done regularly

Lupus medicines:

Help your doctor and yourself

- Keep a list of medicines you currently take
- Give it to your doctor every visit
- Keep a list of medicines tried in the past and why they were used and why they were stopped
- Always go to the same pharmacy

Lupus: Life Expectancy

- Before steroids (before 1949)
 - 83 % died within 2 years of diagnosis
- After steroids available
 - (1964) 69% alive at 5 years
- Current (1999 - 2005) :
 - 86 - 93% alive at 5 years
 - Most lupus patients, no major organ involvement most likely have a normal life expectancy

What kills lupus patients?

- Early after diagnosis (1st 1-2 years):
 - Active lupus disease
 - Severe infections
- Later after diagnosis
 - Cardiovascular disease
 - 1/4 - 1/3 of all lupus patient deaths
 - Heart attacks and strokes
 - Blood clots
 - Severe infections

Preventing Infections:

- Use therapies that don't suppress immune system
- Seek treatment for infections ASAP
- Vaccinations
- Prophylactic antibiotics

Preventing Infections:

- Use therapies that don't suppress immune system
(Decreases the need for immunosuppressant medicines)
 - Use **sunscreen** daily
 - Avoid the sun
 - Take **Plaquenil** daily
 - Take your **vitamin D** supplement faithfully
 - Do not smoke
 - Proper rest
 - Regular exercise
 - Eat healthy

Preventing Infections:

■ Vaccinations

■ Pneumovax

- Pneumonia = most common cause of death from infection in lupus
- Prevents pneumonia from pneumococcus bacteria
- 2-3 times in life (5 years apart)

■ Prevnar

- Life time pneumonia shot

■ Flu shot (influenza vaccine)

- Kills 30,000 – 50,000 Americans yearly
 - Majority are immunosuppressed, elderly, and unvaccinated
- Every summer/fall
- Never causes the flu

Special Vaccine Considerations:

- **Avoid live vaccines** (if on immunosuppressant medicines):
 - MMR (measles, mumps, rubella)
 - OPV (polio) (OK to get eIPV)
 - BCG
 - Vaccinia
 - Typhoid
 - Yellow fever
 - Nasal influenza vaccine (FluMist)
 - **Shingles (Zostavax) (ask your doctor)**

Special Vaccine Considerations:

- Do not get any vaccination if:
 - Lupus is significantly active
 - On 40mg a day of prednisone or more
 - Wait until you are on 20mg a day or less

Treating infection

- Call and see your **primary care doctor** (or go to ER) ASAP if:
 - Have a fever
 - Shaking chills
 - Coughing up sputum
 - Facial pain and fever or drainage
 - Urination pain, frequency, urgency

Treating Infection:

- Always list sulfa as an allergy!

Lupus:

Avoiding heart attacks

- Heart disease
 - This is the #1 cause of death in lupus patients
 - Lupus patients get heart attacks at a younger age
 - Steps to prevent:
 - Eat correctly (Mediterranean diet)
 - Exercise regularly
 - Don't smoke
 - Work with your primary care physician very hard to:
 - Lower cholesterol
 - Normalize blood pressure < 135/85
 - Prevent/treat diabetes

Lupus:

Avoiding Cancer

- Cancer = #4 cause of death
- Lupus patients get cancer more often
- Cigarettes increases lung cancer
- Human Papilloma Virus Cancers Increased:
 - Anal, Cervical, Vaginal, Skin, Mouth
- Prevention:
 - Cancer screenings with PCP
 - Gardasil (HPV vaccine)
 - No smoking!

Conclusion

- All SLE patients should take Plaquenil
- Use sunscreen every day
- Take your vitamin D faithfully
- Get Pneumovax, Prevnar, and flu vaccines
- Have infections treated ASAP
- Never smoke!
- Control risk factors for heart disease

Conclusion

- How can you live a long, normal life with SLE?
 - Learn everything you can
 - **Lupus Foundation of America**
 - Read and do “**The Lupus Secrets**” by Don Thomas, MD
 - Books (**The Lupus Encyclopedia** by Don Thomas, MD)
 - Be compliant with therapy
 - Be compliant with doctor visits
 - Take charge of your health

Take Charge of Your Life!

