

# 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

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St. Martin  
cured  
Bishop of  
Liege of  
lupus  
10<sup>th</sup> 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

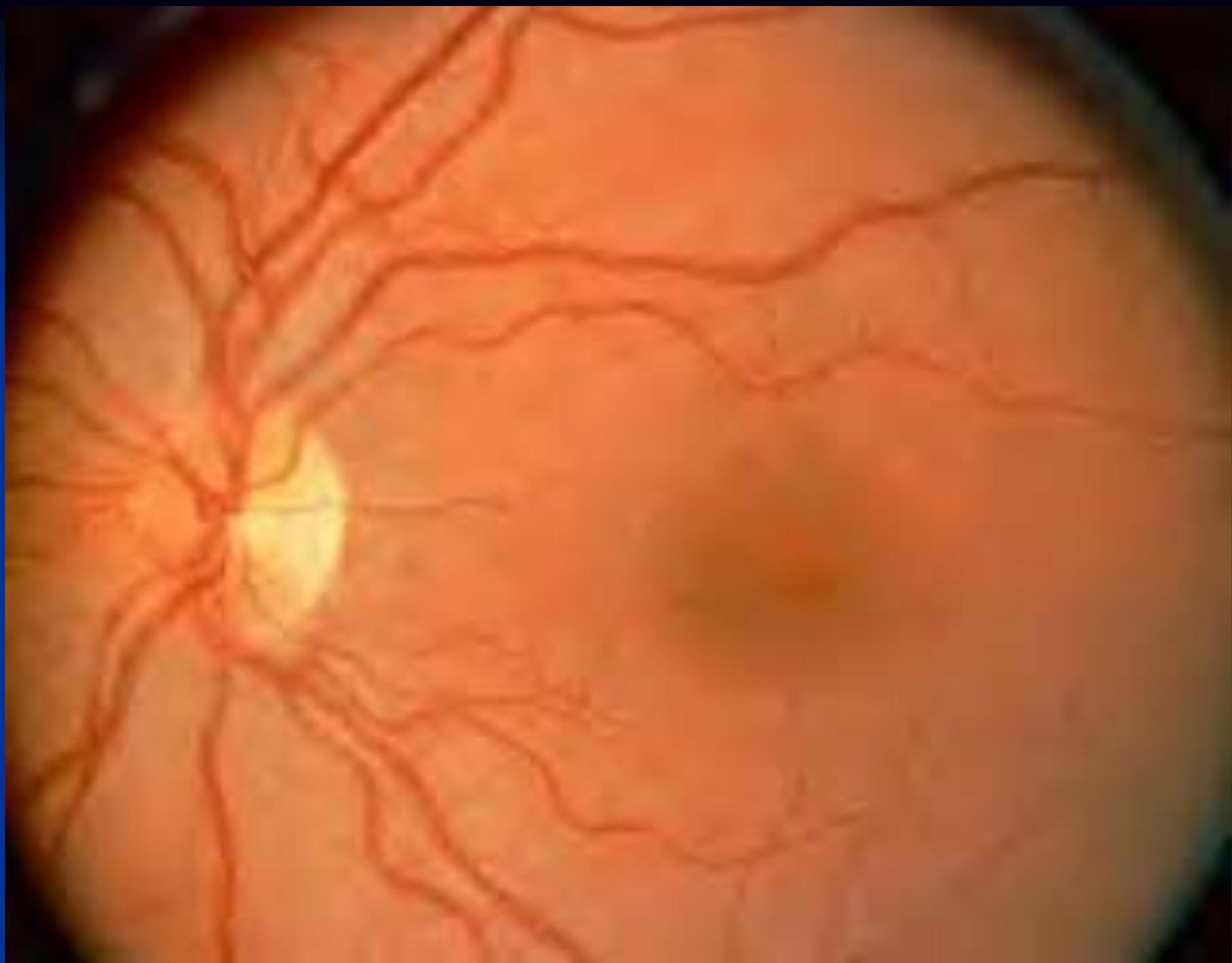
# Plaquenil and Chloroquine: Eye damage!

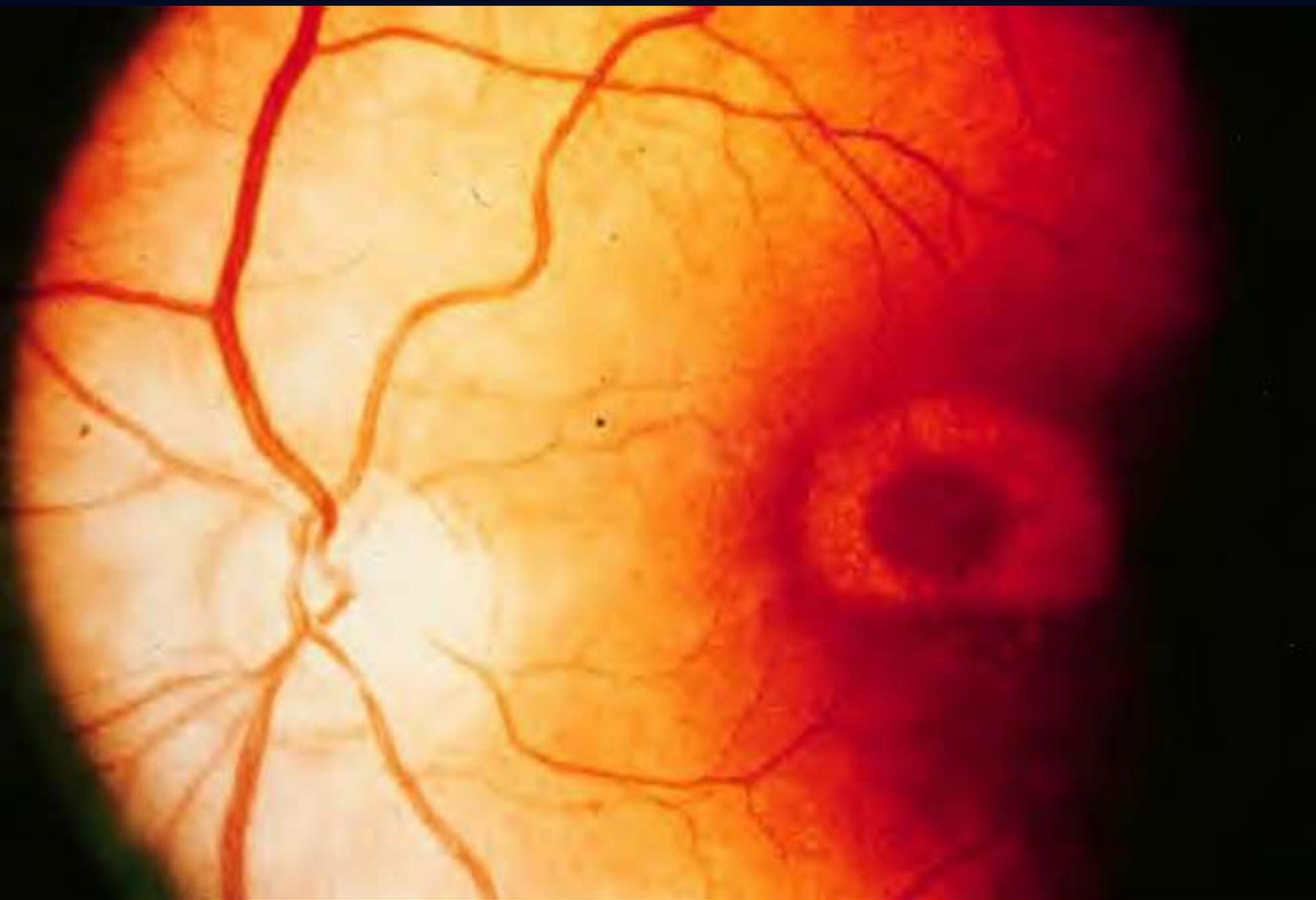
- The medicine can deposit on the back of the eye (retina)
- Causing vision loss
- Uncommon with Plaquenil
- More common with Chloroquine
  - 10% of users

# 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 1<sup>st</sup> 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 (1<sup>st</sup> 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!

