Systemic Lupus Erythematosus
The Basics.

Jagindra .N. Mangru, M.D.
Cumming Rheumatology and Arthritis, LLC.
960 Sanders Road, Ste. 700
Cumming, GA 30041
770-887-5159 (P), 770-887-9496 (F)
www.cummingrheumatology.com
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What is LUPUS?

• Systemic lupus erythematosus (also called SLE or lupus) is a chronic inflammatory disease that can affect the skin, joints, serosa, kidneys, lungs, nervous system, and/or other organs of the body.

• Autoimmune – When the immune system begins to react against the body’s own tissues.
EPIDEMIOLOGY.

- Young women with peak incidence 15-40 years (child bearing).
- 1.5 million in the US.
- Prevalence of 0.5%, however varies with race and socioeconomic status.
- Familial aggregation.
- Most cases are sporadic.
What causes Lupus?

**Innate Susceptibility**
- HLA type (DR3/2)
- Immunoregulatory genes (multiple)
- Complement levels
- Hormonal levels

**Environmental Stimuli**
- UV exposure
- Microbial response
- Drugs

**Autoimmune Proliferation**
- Hyperactive B-cell/T-cell activation
- High ratio of CD4:CD8 T-cells
- Defective immune complex clearance
- Impaired tolerance

**Autoantibody Production**
- Apoptosis & self-exposure
- Self-recognition
- Foreign-Ab cross-reaction
TYPES OF LUPUS.

• Systemic.
• Cutaneous.
• Drug Induced.
• Neonatal.
The most common signs and symptoms include:

- Fatigue and fever
- Joint pain, stiffness and swelling
- Butterfly-shaped rash on the face that covers the cheeks and bridge of the nose
- Skin lesions that appear or worsen with sun exposure
- Fingers and toes that turn white or blue when exposed to cold or during stressful periods (Raynaud's phenomenon)
- Shortness of breath
- Chest pain
- Dry eyes
- Headaches, confusion, memory loss
Symptoms of LUPUS

- Serositis – chest, abdominal, cardiac pain.
- Oral ulcers – painless.
- Arthritis – joint pain and swelling.
- Photosensitivity – rash with sun exposure.
Symptoms of LUPUS

• Blood dyscrasias – anemia, leucopenia, thrombocytopenia.
• Renal dysfunction – proteinuria, casts.
• Autoimmune - +ANA
• Immunologic – SSA, DsDNA, Smith antibody, Anti-cardiolipin antibodies.
• Neurologic – headaches, depression, seizures, psychosis.
Symptoms of LUPUS

• Malar rash
• Discoid rash.
TRUE or FALSE

- A positive ANA means I have LUPUS.

FALSE

FALSE
Diagnostic Criteria

- 4/11.
- 3/10 with + ANA.
- + biopsy.
Complications of LUPUS.

- Cardiovascular disease.
- Infections.
- Renal Failure.
- Vasculitis.
- Drug Effects – cancer, AVN, Osteoporosis.
- Pregnancy related – Hypertension, Lupus flare, kidney disease, fetal loss.
Treatment of LUPUS

Treatments are symptom specific

• PREVENTION:
  - Careful and frequent medical evaluation.
  - Avoid direct exposure to sunlight, Echinacea, drugs – Hydralazine, Sulfa, TNF inhibitors.
  - Use a sunscreen with at least SPF 30.
  - Screening with SS-A antibody.
  - Early pre-natal care.
  - Use of anti-oxidants – Vitamin E, Fish Oil, Flaxseed, Vitamin D.
  - Avoid smoking.
Treatment of LUPUS

- NSAIDS (Naproxen).
- Anti-malarials.
- Corticosteroids (Prednisone).
- Cyclophosphamide (Cytoxan).
- Methotrexate.
- Azathioprine (Imuran).
- Cyclosporine.
- Mycophenolate Mofetil (Cellcept).
Treatment of LUPUS

• Modification of the immune system:
  - Rituximab (anti-CD 20 antibody)
  - Benlysta (BLyS inhibitor)

• Stem Cell Transplantation
Treatment of LUPUS

• Education.
• LUPUS Support – Lupus Foundation.
• Learning to live with a chronic disease
  - praying, meditation
  - stress reduction
  - behavior and lifestyle modification
SUMMARY

• Lupus is an autoimmune disease affecting multiple organ systems.
• Multiple factors contribute to disease expression.
• Treatment options are multi-factorial.