

# Understanding Systemic Lupus Erythematosus in Children and Young Adults

Training and Education for School Healthcare Providers



AMERICAN COLLEGE  
of RHEUMATOLOGY  
*Empowering Rheumatology Professionals*

# Welcome.

We are excited to speak with you today about lupus.

# Before we get started...

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# Learning objectives

After this presentation, you should:



Recognize the signs and symptoms of lupus and know what to do if lupus is suspected



Understand the general principles of lupus treatment and how it can affect students



Identify which populations have a higher risk of developing lupus



Describe how a school healthcare provider can support a student with lupus



Be familiar with the Lupus Care Plan and the Lupus Transition Plan for a student with lupus

# What Is lupus?

# What is an autoimmune disease?

The immune system is designed to attack infections.

**Autoimmune diseases** occur when the immune system mounts a response against one's own body.

Lupus is one type of autoimmune disease.

# So what is systemic lupus erythematosus (SLE)?

Lupus is an inflammatory, multi-system, autoimmune disease of unknown cause.

Lupus can be mild to severe or life-threatening, or anything in between.

The diversity of clinical symptoms is great, and all organ systems are vulnerable.

Lupus is NOT contagious. You cannot “catch” lupus.

There is no cure but treatments are available.

Lupus is characterized by periods of flare and remission and can culminate in irreversible, end-stage organ damage.

# Multiple factors contribute to lupus

Genetics

Race

Age

Estrogen

Sunlight

Certain medications

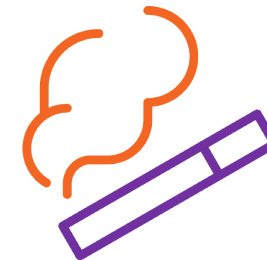
Infection

Smoking

Stress

Toxins

Other unknown factors



# Who is most impacted by lupus?

# Systemic Lupus Erythematosus in Adulthood

**Prevalence** 3.3 – 24 per 100,000 worldwide

**Incidence** 0.3 – 2.2 per 100,000

Hispanic, Asians, and Native Americans are 2-3 times **more likely** to develop lupus than Caucasians.

African American women have **two to three times** higher risk than white women.

People with **lower incomes** are less likely to receive recommended care, and poverty is associated with poor outcomes.

African American, Hispanic / Latino, Asian, and AI/AN women have the **highest prevalence**.



# Childhood Onset Systemic Lupus Erythematosus

**Prevalence** 1.89 – 25.7 per 100,000 persons

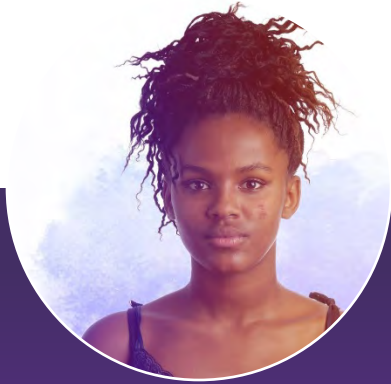
**Incidence** 0.36 – 2.5 per 100,000 per year

Female children are **at least 4 times more likely** to develop lupus than male children.

African American, Hispanic / Latino, Asian, and AI/AN children have higher **prevalence** than white children.



# Impact of Lupus on Children



15-20 percent of lupus patients will present in their childhood years.



Children and adolescents often have more severe disease at onset than adults.



Children with lupus are also more likely to have kidney and neurologic disease.

You should know that lupus impacts African American and Hispanic / Latino women **at a younger age** and more severely than white women.

These populations are also more likely to have major organ involvement.



# Children who are diagnosed with lupus face additional challenges outside of the lupus itself:



## Psychosocial impact

- » Missed school
- » Decreased extracurricular activity due to unpredictable symptoms
- » Fatigue
- » Feeling poorly
- » Social isolation due to being different from peers



## Adverse effects from medications

- » Growth delay
- » Early osteoporosis
- » Changes in appearance
- » Other complications

# The challenges and importance of diagnosing lupus

A lupus diagnosis can take as long as **two or more years** and include visits to three or more health care providers.



# Why is the diagnosis of lupus so challenging?



## LUPUS IS:

- » “the great imitator”
- » can mimic other conditions like viral syndromes, malignancies, allergic reactions, and stress
- » sometimes associated with depression or pain syndromes such as fibromyalgia

## SYMPTOMS MAY BE VAGUE, INCLUDING:

- » fatigue
- » achiness
- » stiffness
- » low-grade fever
- » swollen lymph nodes
- » rashes

## SYMPTOMS MAY:

- » develop slowly
- » come on suddenly



This delay in diagnosis can be **devastating** for a person with lupus.

### **THIS COULD LEAD TO:**


- » organ failure
- » a five-fold increased risk of death

With an early diagnosis, the chances of a person with lupus living a full life with a manageable, chronic disease are increased.



Specific racial and ethnic minorities with lupus have higher mortality rates than white individuals. The reasons for these are still not fully understood. Independent of race/ethnicity, it is thought that lower socio-economic status, and impaired access to specialists to diagnose lupus early contributes to this higher mortality risk. In addition, there may be biologic and other factors which also contribute to this risk.





Diagnosis is best confirmed by an experienced clinician, specifically a rheumatologist, a dermatologist, and/or a nephrologist. They will use symptoms, physical exam, lab tests, and possibly imaging or other studies to confirm the diagnosis.

Let's review some of the symptoms and manifestations of lupus.

# Lupus on the Outside



Joint swelling



Malar rash



Mouth ulcers



Raynaud's Phenomenon



Discoid rash



Vasculitis



Alopecia

It is also a **photosensitive disease**, which means sunscreen and other sun protection is a must!



# Lupus on the Inside



Achiness, headache



Fatigue



Memory loss / brain fog



Depression

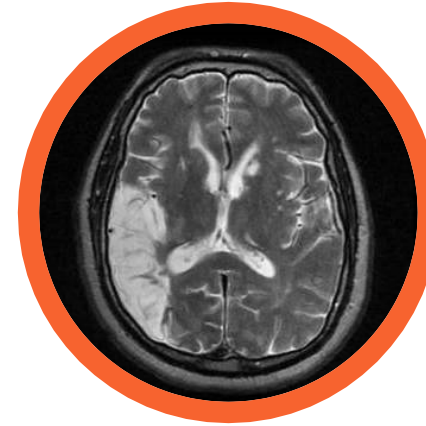
# Illustrations of Organs Impacted by Lupus



Serositis



Pericardial effusion



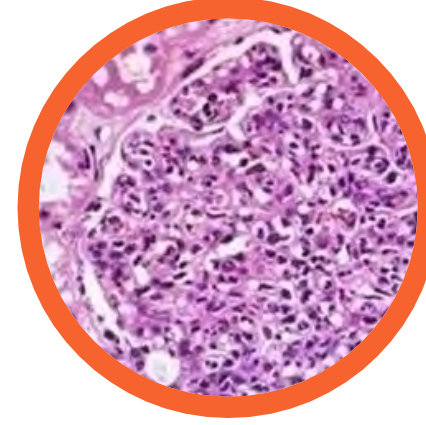
Stroke



Brain atrophy



Spherocytes



Glomerulonephritis

# Lupus manifestations

## MOUTH

- Oral ulcers

## LUNGS / HEART

- Serositis

## KIDNEYS

- Proteinuria
- Hematuria

## MUSCLE & JOINTS

- Arthritis
- Myositis

## BRAIN

- Seizures
- Psychosis

## SKIN

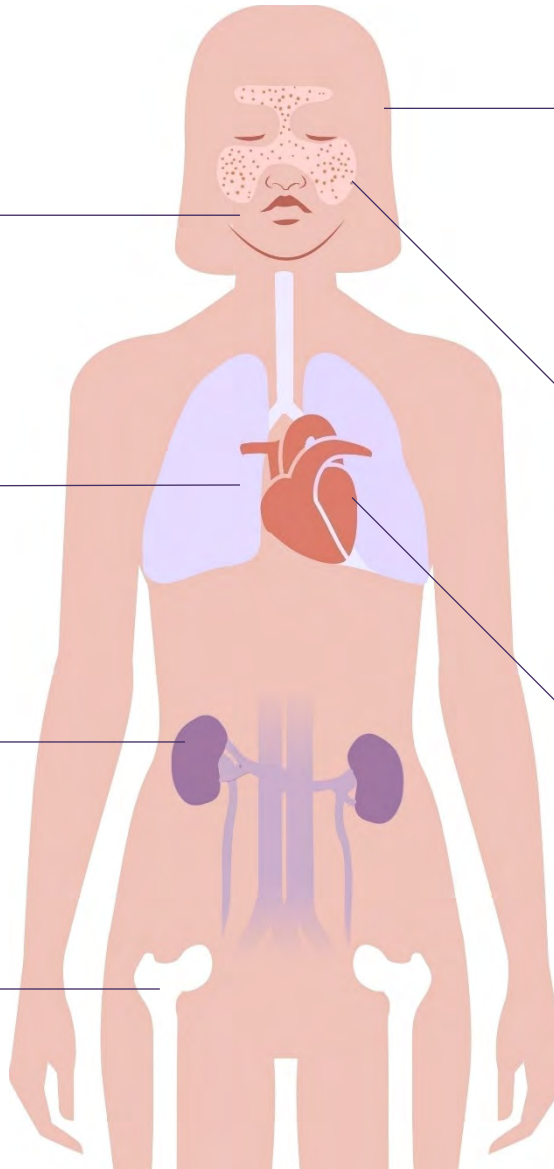
- Malar rash
- Discoid lesions
- Photosensitivity

## BLOOD

- Low blood count

## IMMUNOLOGIC

- Immunologic disorder
- Antinuclear antibodies (ANA)



# Lupus Flares

Flares manifest in a variety of ways



- › Increased pain and fatigue
- › New organ system involvement
- › Rashes
- › Worsening of pre-existing symptoms

Many potential triggers for lupus flares



- › Sunlight
- › Certain medications (sulfa antibiotics)
- › Infections

How do you treat lupus?

# General Principles of Treatment

Immunosuppressive/Immunomodulatory medications

Most treatments aim at getting inflammation under control and decreasing excessive immune response

Most patients require daily medications for ongoing management, but some may require steroids during severe flares and IV infusion



# Treatment

## Daily Medications:

Hydroxychloroquine (Plaquenil),  
Mycophenolate Mofetil/Mycophenolic Acid  
(CellCept/Myfortic),  
Azathioprine (Imuran),  
Methotrexate



## IV Infusions

Belimumab (Benlysta), Cyclophosphamide,  
Rituximab, IVIG



## Steroids

Prednisone or IV steroids for severe flares or  
specific organ involvement/endangerment

Some patients may need to be on prednisone  
for months or years



## NSAIDs

Avoid in kidney disease  
Helpful for MSK involvement (joint pain and  
arthritis)

# Treatment – Side Effects

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Increased risk for serious infections

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GI symptoms/GI intolerance (diarrhea, nausea)

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Infertility/decreased fertility

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Weight gain

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Mood disturbance

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Acne

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Adverse effects on bone metabolism

How can a school healthcare provider help?

# General support

- » Talk openly about lupus and its impact
- » Help combat the stigma by talking about challenges the student may face daily
  - › i.e., changes in physical appearance due to rashes, hair loss, weight gain (from medications); changes in mood, ability to concentrate
  - › Students may feel isolated due to missing school for appointments, treatments, illness
- » Understand physical limitations from joint pain and fatigue that can cause difficulties in participating in physical activity
  - › Limitations may be temporary and should be based on the student's disease activity
- » Ask the student directly what they need from you as a school healthcare provider. Each individual experiences symptoms and challenges differently.

# Day-to-day support

- » Help students apply sunscreen daily and as needed outside
- » Encourage students to:
  - › Get adequate rest
  - › Know their medications and take them regularly (keep a list on their phone)
  - › Live life normally with school activities, healthy diet, and exercise
- » Keep an open line of communication with the student
- » Have students speak to a counselor or mental health professional if they feel sad or depressed
- » Watch for signs of bullying
- » Recognize May as Lupus Awareness Month and use the opportunity to educate others

# Empowering Students with Lupus

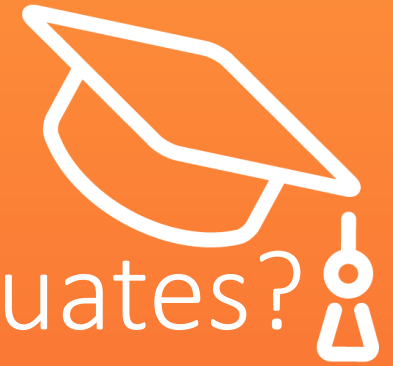
- » Encourage the student to communicate about any hospitalizations and/or doctor's visits that require school absence.
- » Encourage the student and caregivers to keep an open line of communication about new symptoms and come up with an action plan to address those symptoms.
- » If the caregivers are agreeable and consent, maintain phone numbers for the student's rheumatologist to discuss any new concerns regarding the student's health.
- » Discuss the Lupus Care Plan with the student and their caregivers to develop a plan of action during symptomatic flares.

# Lupus Care Plan

- » The Lupus Care Plan is designed for use by the school healthcare provider to identify symptoms of lupus and how to manage those symptoms in the school setting.
- » The plan is to be used by the school healthcare provider to:
  - › Outline management of mild to moderate/severe symptoms, parameters, and procedures for the school healthcare provider to follow during a lupus flare
  - › Address the psychosocial and physical changes that can occur with lupus as well as the treatment of lupus
- » Allows for open conversation between the provider, caregivers, and school healthcare provider to discuss the student's lupus symptoms and how to manage their lupus at school.



What happens when the student graduates?



# Transition from Pediatric to Adult Care

- » Vulnerable time period
  - » Risk for progression to end stage renal disease, risk of death due to gaps in care
  - » Potential for unscheduled emergency room visits, hospitalizations
  - » Higher risk groups: public insurance, history of CPS involvement, recent ER visits/admissions
- » Period of increased responsibility, desire for autonomy
- » Differences in adult and pediatric clinic visits
- » Transitioning care is a *process* over many years, not a single point in time



# Transition of Care

Our transition of care package is intended to outline the process and help assure a seamless transition to independence and self-management.

School healthcare providers can assist by helping students complete their transition plan and transition readiness assessments, and opening dialogue about the process.

Smooth transition is essential for care continuity and continued control of their disease.

During this time, they must learn to manage their disease without direct supervision from a caregiver.







Children “age out” of the pediatric healthcare system in their late teens/early 20s.

# Transition of Care Plan

- » The Transition of Care Plan is:
  - › To be used by a school healthcare provider to assist a student begin to assume independent management of his/her disease without direct supervision from a caregiver
  - › To be developed during throughout high school to enable a student to learn to independently make doctor appointments, refill prescriptions, and know how to access the health care system by the end of their senior year.
- » School healthcare providers can assist in this process by helping students complete their transition plan, transition readiness assessments, and opening dialogue about the process.



# In summary

-  Early symptoms can be non-specific and be easily mistaken for other illnesses or syndromes.
-  It is a chronic disease that can be difficult and take a long time to diagnose.
-  Symptoms may be severe or mild. They can also be transient or prolonged and independent of one another. Treatment is covered in the Lupus Care Plan.
-  90 percent of lupus patients are women between ages 15-45. It is 2-3x more common in people of color.
-  It can affect 11 organs and potentially lead to irreversible damage.
-  Lupus affects up to 1.5 million people across the United States

# Working together to take care of a student with lupus



A team approach can be very helpful in taking care of a child or adolescent with lupus.



People from different cultural and socioeconomic backgrounds experience illness and treatment differently.



A school healthcare provider is in a unique position to provide support to a student that has been diagnosed with lupus.



Through education and planning, we can empower youth with lupus to successfully navigate the transition to adulthood.

# Resources and Information

Ongoing care for people with lupus is a team effort, and it is a matter of life and death.

For presentations, videos, interactive case studies and CE/CME courses, visit The Lupus Initiative at <http://tlitools.org/lectures>.

Thank you.

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