



Diagnosing Lupus in Pediatric Patients



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What is Lupus?

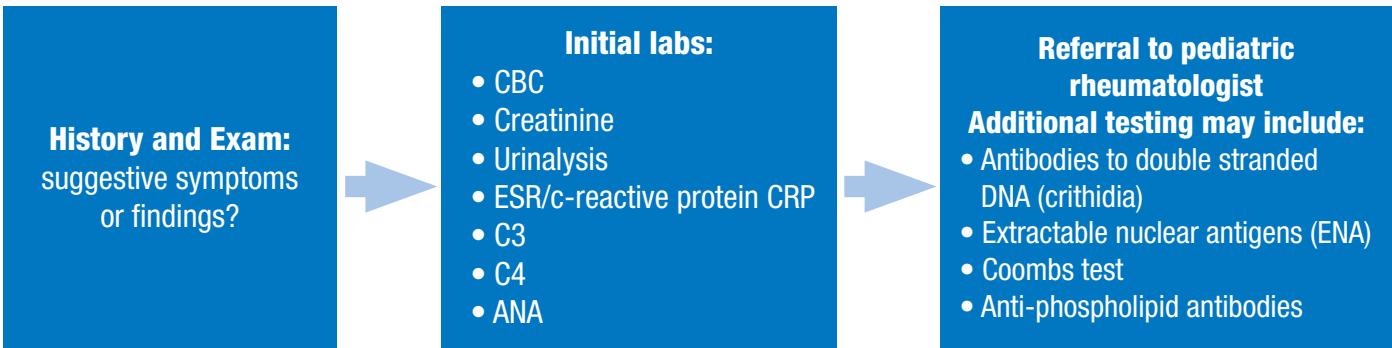
Systemic lupus erythematosus (SLE; lupus) is a chronic and heterogeneous autoimmune disorder that can affect any organ in the body. The disease is more severe in children and adolescents compared to adults. Uncontrolled lupus can cause severe complications including kidney damage from lupus nephritis and psychosis, seizures, stroke and cognitive problems from central nervous system involvement. Early diagnosis is important to prevent organ damage. Lupus can be confused with other conditions that have similar symptoms including infections, leukemia, other types of arthritis such as juvenile idiopathic arthritis, and other autoimmune diseases. Pediatric lupus has a prevalence of 3.3-8.8 per 100,000 children and is more frequently observed in certain ethnic backgrounds and less common in caucasian children.

Symptoms of Lupus

Children and adolescents may present with a variety of symptoms of lupus, several of which may also be shared with other autoimmune diseases. Due to the variable manifestations, a high index of suspicion and thoughtful approach to screening is needed to avoid unnecessary delays in diagnosis. Most physicians use the 11 criteria for Lupus provided by the American College of Rheumatology (ACR). Usually, at least 4 of the 11 ACR criteria should be present to make a diagnosis of SLE.

SYMPTOMS OF LUPUS	
Unexplained fevers Unexplained weight loss Enlarged lymph nodes Alopecia Rashes	Unexplained high blood pressure Sores in mouth or nose Joint pain and swelling Chest pain Shortness of breath Kidney disorder Abnormalities in the blood

Diagnosing Lupus



What Laboratory Tests Help to Diagnose Lupus?

A multisystem history and exam are critical to identify symptoms and findings that suggest lupus. The next step is to look for other laboratory findings that suggest lupus. There are many types of laboratory tests available, however, not all of them are equal in their performance, which is why it is useful for physicians to discuss with laboratory scientists and diagnostic laboratory staff the method of testing used, and its analytical characteristics, including limitations.

While an anti-nuclear antibody (ANA) test is positive in 99 percent of individuals with lupus, approximately 15-20 percent healthy individuals have a positive ANA test. Thus, the ANA test by itself is a poor screening test for lupus (many false positives; sensitivity >95% but specificity <40%). If the history, exam plus and the laboratory testing are suggestive of lupus, additional laboratory testing can be performed to include a direct Coombs test, antiphospholipid antibodies, antibodies to double stranded DNA (also known as *Crithidia* test) and antibodies to extractable nuclear antigens (ENA): anti-Smith, anti-SSA, anti-SSB, anti-ribonuclear protein (RNP). The differential diagnosis of SLE is broad and can include infections, malignancy and other autoimmune conditions. It is important to correlate laboratory testing with clinical context.

LABORATORY FINDINGS THAT SUGGEST LUPUS	
Low white blood cell count Anemia Low platelet count Urinalysis with protein, unexplained red or white cells	Elevated serum creatinine Elevated ESR or c-reactive protein CRP Low C3 and/or C4 Positive anti-nuclear antibody (ANA)

The following tables list the tests that are recommended for diagnosis and referral of patients with suspected lupus. Ordering automatic reflex testing can save patients from additional visits and blood draws, as well as ensure the rheumatologist has complete information.

TEST ID	REPORTING NAME	AVAILABLE SEPARATELY	ALWAYS PERFORMED
ANA2	Antinuclear AB, S	Yes	Yes
CCP	Cyclic Citrullinated Peptide Ab, S	Yes	Yes
IM_01	Interpretation	No	Yes

TEST ID	REPORTING NAME	AVAILABLE SEPARATELY	ALWAYS PERFORMED
CMA	Centromere Ab, IgG, S	Yes	No
CASMT	ANA2 Cascade	No	No
RIB	Ribosome P ab, IgG, S	Yes	No
ENAE	Ab to Extractable Nuclear Ag Eval, S	Yes	No
ADNAR	dsDNA Ab with Reflex, IgG, S	Yes	No

If Findings Are Consistent With Lupus, Refer to Pediatric Rheumatology

A pediatric rheumatologist can confirm the diagnosis by evaluating the history, examination and laboratory findings. Further laboratory testing may be performed. Additional testing, including tissue biopsy, depending on affected organ or specific presentation, may be required.

Lupus is not curable, but lifelong treatment can help prevent end-organ damage. Management and treatment of pediatric lupus requires a multidisciplinary approach. Pharmacological treatment is tailored to severity and extent of disease. Most treatment regimens involve some form of immunosuppression, though mild symptoms can be managed with hydroxychloroquine or related drugs. Non-steroidal anti-inflammatory drugs are used for musculoskeletal pain and arthritis. Selection of immunosuppression and more complex therapies is dependent on the clinical features, severity and progression of disease.

Lupus Clinic at Nationwide Children's Hospital

The Nationwide Children's Lupus Clinic provides multidisciplinary care to children and adolescents with lupus. Our team includes rheumatologist, nephrologists, psychologist, social worker and clinical pharmacist. To refer a patient to Rheumatology at Nationwide Children's, visit NationwideChildrens.org/Rheumatology.

Referrals and Consultations

Online: [NationwideChildrens.org/Rheumatology](https://www.NationwideChildrens.org/Rheumatology)

Phone: (614) 722-5525 | Fax: (614) 722-3194

Physician Direct Connect Line for 24-hour urgent physician consultations:
(614) 355-0221 or (877) 355-0221.

Laboratory Testing and Pathology Consultations

Online: [NationwideChildrens.org/Lab](https://www.NationwideChildrens.org/Lab)

Phone: (614) 722-5477 or (877) 934-6575

