Valley Children's HEALTHCARE

Central California
Pediatrics

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Specialty information for physicians who treat children and expectant mothers.

Systemic Lupus Erythematosus in Children: When to Consider the Disease?

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Systemic lupus erythematosus, also referred to as SLE or "lupus," is a chronic, multisystem autoimmune disease. This diagnosis most commonly affects young women, but can affect children and cause childhood-onset lupus (cSLE).

The diagnosis of lupus can be challenging as the clinical features vary considerably. SLE can present with non-specific symptoms such as fatigue, fever and weight loss to fulminant, life-threatening disease. Initial evaluation for SLE requires a careful history and physical exam, along with laboratory testing to identify features that are characteristic of SLE.

Children and adolescents with lupus may present with the following features:

- **Musculoskeletal disease:** Arthritis and arthralgias occur in > 90% of patients and are often the initial presenting symptom of SLE.
- **Skin lesions:** Include malar rash, discoid lupus and photosensitivity. The malar rash is an erythematous rash distributed in a butterfly pattern over the nose and cheeks, but characteristically spares the nasolabial folds.
- **Oral and nasal ulcers** that are often painless.
- **Renal disease:** May present as acute or chronic renal failure. Acute nephritic disease is encountered in more than 50% of patients with SLE and is an important cause of morbidity and mortality.
- **Neuropsychiatric involvement of SLE:** Consists of a broad range of neurologic and psychiatric manifestations, including cognitive dysfunction, organic brain syndromes, delirium, psychosis, seizures, headache and/or peripheral neuropathies.
- **Hematologic abnormalities:** Includes autoimmune hemolytic anemia, leukopenia and thrombocytopenia.
- **Arterial or venous thrombosis** can occur in patients with antiphospholipid syndrome.
- **Pulmonary involvement** manifests as pleurisy, pleural effusion, pneumonitis, pulmonary hypertension or interstitial lung disease.
- **Cardiac disease due to SLE:** May present as pericarditis or myocarditis.

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If Systemic Lupus Erythematosus is Suspected

If SLE is suspected, a complete blood count and differential, as well as serum creatinine level and urinalysis, should be obtained. Other laboratory tests that support the diagnosis of SLE, if abnormal, include: antinuclear antibodies (ANA), anti-double-stranded DNA, anti-Smith, antiphospholipid antibodies, C3 and C4 or CH50 complement levels, erythrocyte sedimentation rate and/or C-reactive protein levels, and urine protein-to-creatinine ratio. It is important to note that ANA tests can be falsely positive and false-positive rate varies from about 3% with ANA titers of 1:320 to about 30% for ANA titers of 1:40 among healthy controls.

The American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR) published new criteria for the classification of SLE in 2019. These criteria require an ANA titer of at least 1:80 in addition to other criteria that include the seven clinical domains (constitutional, hematological, neuropsychiatric, mucocutaneous, serosal, musculoskeletal, renal) and three immunologic domains (antiphospholipid antibodies, complement proteins, SLE-specific antibodies). Labs are ordered based on symptomatology and the degree of illness.

If lupus is suspected, initial labs should include complete blood count with differential, comprehensive metabolic panel, inflammatory markers including ESR and CRP, urine analysis to look for proteinuria and hematuria, complements including C3 and C4, antinuclear antibody testing with immune-fluorescent assay and double-stranded DNA.

Management of SLE often depends on the individual patient's disease severity and the treatment goals are to suppress the immune system and prevent permanent organ damage. Prompt diagnosis and referral to a rheumatologist with experience can help mitigate the morbidity and mortality associated with the disease.

Medical Staff News

The following pediatric specialists recently joined Valley Children's:

Emergency Medicine
Melissa Hersh, MD

Gastroenterology
Ayesha Baig, MD

Pathology
Amy Davis, MD

Pulmonology
Modesto - Pelandale Specialty Care Center
Sebnem Ozdogan, MD

Pediatric Clinical Symposium Virtual Education Series

Saturday 2 Hour Events
9 - 11 a.m.

Neuroscience Feature on Pediatric Epilepsy
Steven Ehrreich, MD
Julia Sharma, MD
Saturday, October 24

Non-Accidental Trauma
John Kinnison, MD
Saturday, November 14

Register today at cmetracker.net/vch or for more information contact programs@valleychildren.org.