UCLA pediatric rheumatologists and pediatric nephrologists see pediatric lupus patients in a convenient combined clinic that provides close coordination of services for patients and families. UCLA’s program focuses on total lupus care beginning with careful diagnosis and close monitoring to control flare-ups and mitigate organ damage.

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by multiple antibodies directed against the body’s own tissues (autoantibodies), resulting in inflammation of blood vessels and connective tissue. Childhood-onset lupus has a prevalence of three to nine cases per 100,000 children. About 80 percent of lupus patients are female, with the median age of onset being 11 to 12 years — often occurring at the start of puberty. The disease impacts a number of organ systems and can cause a variety of symptoms.

While disease manifestations can be unpredictable, initial symptoms usually include rash, fever, arthritis, fatigue, anorexia and weight loss. A defining characteristic of lupus, occurring in about 60 to 75 percent of cases, is the malar, or butterfly, rash across a child’s cheeks and nose. Compared to adult-onset cases, childhood-onset lupus tends to produce a more severe clinical course including involvement of the kidneys and heart. Renal disease occurs in 50 to 75 percent of all childhood-onset lupus patients, often within two years following diagnosis.

“Lupus has been very difficult to understand,” says Deborah McCurdy, MD, professor of pediatric rheumatology. “It is not a single-gene disease, and probably does not involve the same genes in all ethnic groups. We don’t know exactly what genes are involved and how they interplay, but in the last two decades there have been major advances in understanding the immune system. We now have some ideas of what genes are turned on and what pathways are activated. We are getting some better ideas on how to suppress those pathways so they are not overactive.”

UCLA’s comprehensive pediatric clinic provides state-of-the-art care that recognizes the evolution in treatments.

“It is a big advantage to patients to have one clinic where doctors in rheumatology, nephrology, cardiology and pulmonology interact. It allows not only for patient convenience, but for a team approach to care,” Dr. McCurdy notes. “Patients don’t end up getting one recommendation from one healthcare provider and another from a different provider. It improves clinical care.”
Evolving diagnostic criteria

A timely and accurate diagnosis is important in limiting adverse outcomes, yet diagnosis can be challenging as lupus tends to mimic other autoimmune diseases and presentation can sometimes be mild and vague, with symptoms resembling influenza. Diagnosis is typically based on fulfilling a number of classification criteria from the American College of Rheumatology (ACR). These criteria are undergoing review and are expected to be refined. UCLA’s lupus experts closely follow updates from the ACR as well as international panels that seek to improve diagnostic criteria.

Patients at UCLA undergo a comprehensive diagnostic workup that includes history, physical examination and laboratory tests to rule out other medical problems, such as infection or malignancy. Patients suspected of lupus undergo an antinuclear antibody (ANA) test. While 98 percent of lupus patients have a positive ANA, which indicates immune system activation, a positive antinuclear antibody test is not definitive, and alone is insufficient to diagnose the disease.

Promising research may soon yield additional biological markers found exclusively in people with lupus.

Careful selection of treatments

Every patient has a unique clinical course, so personalized and comprehensive care is paramount to optimal outcomes. Patients with milder manifestations, such as rash and joint problems, require less aggressive treatment. Treatment can include non-steroidal anti-inflammatory drugs (NSAIDs) and anti-malarial medications.

However, since many pediatric patients have kidney involvement, more aggressive treatment is typically indicated, including use of corticosteroids (oral and intravenous) and immunosuppressive therapies. The first new medication for lupus in more than 50 years, belimumab, was approved by the Food and Drug Administration in 2011. The drug, a human monoclonal antibody that inhibits B-cell activity, is approved for adults but has shown positive results in recent pediatric clinical trials.

Pediatric lupus patients at UCLA may be candidates for one of the numerous clinical trials that are part of the adult lupus program, including research on genetic variations of the disease. The pediatric lupus program is aligned with organizations that advance new treatments, including the Childhood Arthritis and Rheumatoid Research Association.

Care in a multidisciplinary setting

UCLA’s convenient clinic, featuring pediatric rheumatologists and pediatric nephrologists, promotes the desired team approach to lupus care and close coordination of services. Cardiology and pulmonology services can also be made available in the same visit.

Children and adolescents can be referred for psychosocial supportive services. Moreover, patients who qualify for California Children’s Services have access to nursing, social work, physical therapy and other services. UCLA’s medical team works closely with the Lupus Foundation, Lupus LA and the Lupus Research Institute to provide additional family and patient education and support.