

Lupus Diagnosis Can Be an Exercise in Suspicion

BY SUSAN LONDON

SEATTLE — The clinical features of lupus in children may be subtle and easily overlooked, Dr. David Sherry said.

Vasculitis, the pathologic hallmark of lupus, can produce a challenging clinical picture with a wide differential diagnosis, noted Dr. Sherry, who is a pediatric rheumatologist at the Children's Hospital of Philadelphia.

"It's such a black hole, a lot of clinicians just don't want to think about it." But clinicians should think about it under certain circumstances, he said.

One is when a child has constitutional



The spotty malar rash associated with lupus can mimic rosacea.

symptoms that persist. "When you have a kid who is sick and they are not getting better—they still have a fever, they are still losing weight, they have an elevated sedimentation rate, and the 'virus' still isn't going away—you need to think maybe they've got a vasculitic condition," he commented at a meeting sponsored by the American Academy of Pediatrics.

Multiorgan disease can also be a tip-off for vasculitis. "Why should a kid be peeing blood and coughing up blood?" he said. "That's two different organs." Seeing an unusual patient for the symptom, such as a teenager with a heart attack, also should raise a suspicion of vasculitis.

Finally, there is the vasculitic rash, which can have a variety of appearances.

In describing the malar rash of lupus,

textbooks often show photos of a vivid, contiguous red rash in the classic butterfly distribution on the cheeks and nose, according to Dr. Sherry. But what is actually seen clinically may instead mimic rosacea, wind chapping, sunburn, or even acne. In addition, in black children, the rash may be subtle and especially hard to identify.

Key features that can help identify a malar rash of lupus include its distribution, typically with crossing over the bridge of the nose and spreading onto the cheeks, and a well-defined border between the affected skin and normal skin. Children with malar rashes usually have other symptoms or clinical findings too.

Additional clues to the presence of lupus can often be found on parts of the body that are easily overlooked on examination, according to Dr. Sherry. For example, children may have a vasculitic rash on their hands or feet, or a painless ulcer on their hard palate. "You need to look up to see the hard palate," he pointed out. "If you look at the back of the throat, you will miss this."

Discoid rash is less common and causes crusts or scabs that scar. "If you lift up these crusts or scabs, you see what's called carpet tacking—little pinpoint spots of bleeding underneath," he said. "Discoid lupus especially likes the helix of the ear, so pay attention to the helix."

Children also may have so-called lupus hairs, which are fragile and break easily.



This lupus patient's rash is largely confined to the nose.



Children with lupus may have a vasculitic rash on their hands.

"You pull on their hair, you get three, four, or five hairs, even if they just brushed it," he explained. The breakage is accompanied by the presence of short hairs resulting from regrowth.

When lupus is first suspected in children, Dr. Sherry recommended that physicians obtain a complete blood cell count, an erythrocyte sedimentation rate (ESR), a C-reactive protein (CRP) level, a urinalysis, a comprehensive metabolic panel, and an antinuclear antibody (ANA) titer. Lupus has the unique property of producing a high ESR and a normal CRP level—unless the child also has an infection.

An ANA panel can be deferred unless suspicion of the disease is high, he said. Related tests should be guided by symptoms, such as rheumatoid factor assessment in a child with pronounced joint symptoms, creatine kinase assessment in a child with muscle weakness, and coagulation studies in a child with deep venous thrombosis.

Dr. Sherry noted that to be classified as having lupus, children must meet at least 4 of the 11 clinical and laboratory criteria of the American College of Rheumatology, of which a positive ANA titer is merely 1.

In fact, he cautioned, 12%-20% of normal children have a positive ANA titer.

If the ANA result is positive but at a titer of only 1:80 or 1:160, the child is unlikely to have lupus; if it is higher, the

ANA panel should be done. "If the panel is negative, you can cool your jets and cool the mom's jets," he said.

Management in children with lupus, in addition to antirheumatic therapy, includes counseling about sun protection because of photosensitivity, antihypertensive therapy when blood pressure is elevated, and attention to calcium and vitamin D status, both because steroid therapy adversely affects bone health and because lower levels of vitamin D have been linked to increased disease activity. "We give these kids calcium and, if they are low in vitamin D, we certainly can give them that too," Dr. Sherry said.

"The outcome of most kids with lupus is certainly not what it used to be," he said. "More than 90% of our kids do very well and have long-term survival."

In fact, "now that we are saving these



A painless oral ulcer of the hard palate can be easily overlooked.

kids, we have to worry about the side effects of treatment." Hence, where possible, treatment strategies have been modified to reduce long-term toxicity.

Importantly, he concluded, as children with lupus increasingly survive into adulthood, their providers will need to be aware of risks related to the disease and its treatment that may emerge over time, including hyperlipidemia, heart attack, and complicated pregnancy.

Dr. Sherry reported that he had no conflicts of interest in association with his presentation. ■

Sunny Climate Kids Not Immune to Vitamin D Insufficiency

BY DIANA MAHONEY

BOSTON — The high prevalence of vitamin D deficiency found in a cohort of healthy children in a sunny Southwestern climate has prompted a call by the study's investigators for generalized routine screening of vitamin D levels among all children.

In a study designed to assess vitamin D levels in children living in a region with year-round sunshine and to compare vitamin D levels in children with vague musculoskeletal pain with those of children without pain, Dr. Elizabeth A. Szalay and her colleagues at the University of New Mexico Hospital in Albuquerque retrospectively studied

the serum 25-hydroxyvitamin D (25[OH]D) levels of 77 healthy children who were seen for musculoskeletal pain but who lacked a concrete diagnosis to explain their pain (pain group). They also prospectively obtained serum 25(OH)D levels from 35 healthy children without pain.

The study included healthy children aged 2-16 years who were freely ambulatory and could play outside as they chose. It excluded children with any endocrinopathy and those taking medications that affect vitamin D metabolism, such as steroids or bisphosphonates, Dr. Szalay said in a poster presented at the annual meeting of the Pediatric Orthopaedic Society of North America.

The study population (mean age, 9 years) included 66 girls and 46 boys, and was primarily Hispanic (59) and white (37). The average 25-hydroxyvitamin D levels for the pain and control groups were not statistically different, at 28 ng/mL and 31 ng/mL, Dr. Szalay reported, nor did the vitamin D levels vary by month of the year.

Based on the collective data, the mean 25(OH)D level was 29 ng/mL. "While there is no consensus on optimal serum vitamin D levels in children, optimal calcium absorption is seen between 40 and 100 ng/mL," Dr. Szalay said. "Vitamin D deficiency is defined by most experts as a [25-hydroxyvitamin D] level less than 20 ng/mL."

Only 13% of the children had vitamin D levels in the optimal range, while 33% had levels from 30 to 39 ng/mL, 35% had levels from 20 to 29 ng/mL, 16% had levels from 10 to 19 ng/mL, and 3% had levels less than 10 ng/mL—the level at which rachitic changes may occur.

"This is an alarming trend, especially because the location—with its Southern latitude—had been thought to be ideal for adequate vitamin D levels," said Dr. Szalay. Despite the abundant sun exposure and lack of chronic illness, "the percentage of children with sub-optimal vitamin D levels was significantly greater than that demonstrated among healthy teens studied in the 2002 Third

National Health and Nutrition Examination Survey."

The findings seem to suggest that modern lifestyles, even among children living in sun-rich regions, may be taking an ever greater toll on pediatric vitamin D levels and indirectly on pediatric bone health, said Dr. Szalay. "Increasing childhood obesity, declining dairy intake, increased time using computers and video games, decreased outdoor activity, and increased sun-screen usage may be contributing factors impairing availability to vitamin D, which is essential for normal calcium metabolism and bone mineralization."

Dr. Szalay reported having no financial conflicts with respect to this presentation. ■