# Childhood Dermatomyositis Is Almost Never Associated With a Cancer Risk

BY SHARON WORCESTER
*Southeast Bureau*

DESTIN, FLA. — Dermatomyositis is by far the most common form of idiopathic myositis in children, accounting for nearly 90% of cases, Dr. Brian Feldman said at the annual Rheumatology on the Beach.

This differs from adult disease in that only about 14% of adult patients with myositis have dermatomyositis, with 11%-14% experiencing cancer-associated myositis, about 25% having overlap disease, and close to 50% having either polymyositis or inclusion body myositis.

Cancer-associated dermatomyositis almost never occurs in children, thus a work-up for underlying malignancy is not necessary in this patient population, said Dr. Feldman, a professor at the University of Toronto.

The disease in children can range from mild to severe, and a papulosquamous heliotrope rash, usually on the extensor surfaces, is typical. Such a rash that occurs over the knuckles is known as Gottron's rash, and this is also characteristic of the disease.

Calcinosis is a common finding in children, unlike in adults. It can be superficial or deep; in a third of patients, scarring lesions can be seen as long-term sequelae of the disease.

Systemic manifestations and cardiac disease can also occur in children with dermatomyositis, but both are more common in adults than in children.

Another finding is periungual erythema resulting from capillary nailfold changes. In fact, the underlying pathology in the disease is capillary vasculopathy and it has been shown that changes in the capillaries in the nailfold reflect disease activity changes, Dr. Feldman said.

In one study, Dr. Feldman showed capillary density in the nailfold as measured by capilloscopy strongly reflected disease activity in 42 children; as disease activity rose, capillary density was reduced, he said. The relationship did not extend to muscle damage.

**Another tool that is useful for following dermatomyositis patients is the 14-point Childhood Myositis Assessment Scale, which can measure patient ability.**

Disability can be measured using the Childhood Health Assessment Questionnaire. Originally developed for evaluating disability in children with arthritis, this tool has also been shown to be a valid and responsive tool in these patients, he said.

Another tool useful for following dermatomyositis patients is the Childhood Myositis Assessment Scale. The 14-point scale is an occupational measure of muscle function that uses a series of maneuvers standardized by a group of experts in the field to measure patient ability.

"It turns out this is a phenomenal tool to follow children with myositis," Dr. Feldman said, noting that he uses it for every patient at every visit.

The availability of such tools led to the idea that it might be possible to predict outcomes in children with dermatomyositis, he added.

In adults, dermatomyositis has been shown to be progressive, with increasing disability each year. In one study, mortality in adults was high at 25% over a 7- to 8-year period.

In children, however, the outcomes are much better. In one study of 65 patients followed for about 7 years, only 1 died, physical function was excellent in most of the children, and educational and vocational achievements were also excellent. A substantial number of patients, however, remained chronically affected, with more than a third continuing to require medication for the disease at 7 years, including 15% who remained on prednisone.

In a recent study (see related story) of an inception cohort of patients followed using the validated tools, Dr. Feldman and his colleagues showed that there are factors—such as persistent Gottron's rash—that can predict disease course and chronicity. Patients with a persistent rash at 3 months had increased time to remission, and those with persistent rash at 12 months were significantly more likely to have a chronic disease course, he said.

The study also showed that in children, disease remission was almost always permanent. ■

# *Gottron's Rash Predicts Juvenile Dermatomyositis*

DESTIN, FLA. — A persistent Gottron's rash is the best predictor of childhood dermatomyositis disease course, Dr. Brian Feldman said at the annual Rheumatology on the Beach.

In a study of 81 children with this inflammatory connective tissue disease who were followed for at least 3 years, a treatment-resistant Gottron's rash at 3 months following diagnosis predicted significantly longer time to remission (84 months vs. 41 months in those with no rash at 3 months).

At 6 months, predictors of remission included persistent Gottron's rash (hazard ratio 0.5) and nail fold abnormalities (hazard ratio 0.6), and at 12 months, a combination of Gottron's rash and weakness strongly predicted a chronic disease course, said Dr. Feldman, professor at the University of Toronto.

Gastrointestinal symptoms at diagnosis predicted significantly shorter time to remission (34 months vs. 84 months). The reason for this remains unclear, but in a poster on this data presented at the annual meeting of the American College of Rheumatology in November, Dr. Feldman and his colleagues noted that specific treatment regimens used early in the course of the disease may have contributed to the findings, which will be analyzed further.

The study also showed 60% of patients had chronic disease, 37% had monocyclic disease, and 3% had polycyclic disease, suggesting in children, remission is permanent. "When it's gone, it's almost always gone, and we can tell our patients that they are likely to stay in remission for the rest of their lives," Dr. Feldman said.

Patients in the study had a mean age of nearly 8 years and a mean follow-up of nearly 6 years. The median time to remission was 4 years. About half remitted within 4½ years, and at 10 years, about one-third still had chronic active disease, he said.

**—Sharon Worcester**

# Just to Be Sure: Overtreat in Suspected Kawasaki Disease

BY NANCY WALSH
*New York Bureau*

NEW YORK — Don't feel guilty about overdiagnosing and overtreating Kawasaki disease, Dr. Jeffrey R. Starke said at a meeting sponsored by the American College of Emergency Physicians.

"If you look at the risk-benefit ratio of treatment versus the complications if we don't treat, it's clear we should err on the side of overtreatment, especially in children younger than 12 months who are at high risk for developing severe coronary artery abnormalities," Dr. Starke said.

These youngest patients also are more likely to present with incomplete Kawasaki disease. This diagnosis should be considered in infants under 6 months with fever for longer than 6 days and unexplained systemic inflammation. An incomplete Kawasaki disease-diagnosis also should be considered in children with fever for over 5 days and two or three, rather than four, of the features of Kawasaki disease (see box).

In such a patient, if the C-reactive protein is 3 mg/dL or greater and/or the erythrocyte sedimentation rate is 40 mm/hour or more, supplemental laboratory criteria should be obtained. If three or more are present, the child should have an echocardiogram and treatment should begin.

If there are fewer than three of the laboratory criteria, an echocardiogram is needed. If the echo shows cardiac abnormalities, treat. If it is negative and the fever abates, disease is unlikely. If fever persists, repeat the echo, said Dr. Starke, vice chairman of pediatrics at Baylor College of Medicine, Houston.

Standard treatment with intravenous immunoglobulin (IVIG), 2 g/kg as a single infusion is quite safe, though expensive, which is another reason to err on the side of treatment. On occasion, treatment can begin before an echocardiogram. Aspirin is routinely given, though a recent review found insufficient evidence supporting this practice (Cochrane Database Syst. Rev. 2006;doi:10.1002/14651858.CD004175.pub2). High doses of 80-100 mg/kg per day should be given initially.

Some continue the high dose for 14 days and then reduce the dose to 3-5 mg/kg per day, while others maintain the high dose only until 24-48 hours after the patient defervesces, and then switch to the low dose for 2 months for antiplatelet effects.

"I don't know of a single infectious disease expert or rheumatologist who doesn't still use aspirin in addition to IVIG in the treatment of Kawasaki disease," Dr. Starke said.

Some data suggest some benefit of adding corticosteroids to treatment. In a recent prospective randomized trial of 178 Kawasaki patients, children on prednisolone with IVIG had a shorter duration of fever and a faster fall in C-reactive protein, but no difference in coronary artery dilation at 1 month (J. Pediatr. 2006;149:336-41).

If the fever persists or returns more than 36 hours after completion of the IVIG infusion, a second dose can be given.

"But be patient. It's very common for kids to continue to run a fever the day after you give the IVIG dose, even as other symptoms are improving," he said. ■

## Clinical Diagnostic Features of Kawasaki Disease

**Diagnostic Criteria**

Fever greater than 39 degrees C for 5 days, plus four of the following:

- Conjunctivitis, usually bulbar and bilateral
- Mucus membrane changes such as redness and cracking
- Rash, either generalized or local
- Enlarged cervical lymph nodes, usually unilateral and nontender
- Peripheral changes such as swelling or peeling

**Incomplete Kawasaki Supplemental Laboratory Criteria**

- Albumin 3 g/dL or less
- Anemia for age
- Elevation of alanine aminotransferase
- Platelets 450,000/$mm^3$ or more after 7 days
- While blood cell count 15,000/$mm^3$ or higher
- Urine white blood cell count 10/HPF or more

Sources: Dr. Starke; Circulation 2004;110:2747-71.