Refining Treatment of Juvenile Dermatomyositis

Newly created consensus protocols can help clinicians determine the best therapy.

BY AMY ROTHMAN SCHONFELD
FROM A MEETING SPONSORED BY NEW YORK UNIVERSITY

NEW YORK – Because juvenile der- matomyositis is a rare disease and its symptoms often differ from those seen in adult dermatomyositis, pediatricians and family physicians may not recognize it and diagnosis can be delayed, Dr. Brian Feldman said.

He described a recent practice survey that found variability in juvenile der- matomyositis (JDM) treatment and, in the absence of randomized controlled trials, urged clinicians to consult newly created consensus protocols developed by rheumatologists and to gather data that may help to optimize treatment and minimize side effects in the future for those with JDM.

Dr. Feldman described a 15-year-old patient who had progressively deteriorated over a 3-year period despite being seen by pediatricians and dermatolo- gists. His initial symptoms included fa- tigue and elevated muscle enzymes, in- cluding aspartate transaminase (AST), alanine transaminase (ALT), lactate de- hydrogenase (LDH), and creatine phosphokinase (CPK).

Within a year, the patient developed a purple skin rash on his hands, elbows, and knees. He later developed Raynaud’s syndrome and muscle weakness with decreased range of motion that interfered with participation in sports and other activities of daily living. Upon ex- amination, he was quite thin with a scaly rash over his knuckles (Gottron’s papules), healing skin ulcerations on his hands (from severe Raynaud’s), periun- gual erythema, and lipodystrophy and erythema of his forearms. Magnetic reso- nance imaging showed acute inflam- mation of his shoulder girdle, said Dr. Feldman, chief of pediatric rheumatol- ogy at the Hospital for Sick Children in Toronto.

Dr. Feldman said that one of the best diagnostic clues for JDM comes from mi- croscopic examination of nail folds. In this case, the patient had tortuous, bushy nail folds, dilated or missing capillaries (capillary density of 3 per mm of nailfold length while normal ranges from 7-11 per mm), and curtilar overgrowth in- dicative of JDM.

For rheumatologists, the different pre- sentation seen with children compared with adults may hinder a correct diag- nosis. For instance, calcinosis is seen much more often in children than adults. Children are less likely to have some of the systemic symptoms, such as fever, poor weight gain, and pulmonary ef- fects, and they almost never have cardiac problems. Unlike adults with dermatomyositis or polymyositis, children appear to have a fourfold increased risk of malign- nancy, very few cases of malignancy have been associated with JDM. Children are more likely to experience dyspho- nia/dysphagia but are as little likely to have arthritic symptoms (around 58% of each group). Myositis-specific antibodies do not seem to play as important a diag- nostic role for children as adults. Children who develop JDM appear to have a better prognosis than adults. Al- though adults with myositis have appre- ciable mortality (about 10%) and pro- gressive disability, findings from Dr. Feldman’s group show that outcomes were often excellent in children. “We have not had a single death from JDM in 30 years,” he said.

About two-thirds of children with JDM have a polycyclic, chronic, unremitting disease course, but about one-third follow a monocylic course and have disease symptoms that last for 2-3 years and then go away, with, or potentially even with- out, treatment. In a very small proportion of these patients, symptoms may return many years later. It is possible to predict which patients will follow a chronic course and which patients will have early remission. In his experience, the risk of persistent disease falls by 15% each month on record (combining global land and ocean av- erage surface temperatures) and the 304th consecutive month with a global temperature above the 20th-cen- tury average, according to the National Oceanic and At- mospheric Administration.

Scientists predict changes already underway will warm the planet at least 2 or 3 degrees Centigrade this century, and perhaps double or triple if humans don’t alter the behaviors that contribute to climate change. Current scientific estimates suggest that an in- crease of 2 degrees Centigrade may be the “tipping point” that triggers irreversible changes (such as a rise in sea level, and methane release from melting permafrost) with un- predictable but severe consequences for the planet and its inhabitants, Dr. Muller said.

Some projected changes already are being seen in the extreme summer heat in western Russia, flooding in Indonesia, and drought in Australia. Scientists have thought that the 1980s and 1990s were associated with a nearly fourfold increase in Carrion’s disease in Peru’s Ancash region.

In light of the CARRA study’s findings of heterogeneity in the treatment of JDM and the absence of randomized controlled trials, a group of 12 pediatric rheumatologists met to study treatments in JDM using a new approach by develop- ing consensus treatment protocols (Arthritis Care Res. 2010;62:219-25).

“While these findings reflect the cur- rent prescribing practices of rheumatol- ogists, there are almost no randomized controlled trials of medications for JDM, Dr. Feldman pointed out.

This reflects in part the rare nature of the disease, its complexity, the difficulty of conducting studies in children, and the high cost of randomized controlled trials. There have been studies using ad- vanced analytic techniques to provide strong comparative data in lieu of a ran- domized controlled trial. One such study by Dr. Feldman and associates (Arthritis Rheum. 2008;59:989-95) showed that controlled trials of medications for JDM, Dr. Feldman pointed out.

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“This is similar to what has been done in pediatric oncology,” said Dr. Feldman, who was one of the participants. “We are hoping physicians throughout the world will take these protocols off the shelf, and by using standardized doses, follow- ing standard protocols, we will be able to accumulate enough evidence over time to know which is the best therapy.”

In brief, the group recommended three protocols for the treatment of patients with moderately severe JDM: pulse IVMP plus MTX; IVMP, MTX, plus IVIG; or oral prednisone plus MTX. The third protocol is the one followed most often at the Hospital for Sick Children. The treatment protocols are not intend- ed as treatment recommendations, al- though it is hoped that a physician will choose to follow the standardized protocol that most closely reflects his or her preferred practice. It is presented as a “first step to allow comparison of dif- ferent approaches to the treatment of JDM,” he said.

Dr. Feldman has done contract re- search with Bayer Healthcare Pharma- ceuticals. He has referenced unlabeled/ unapproved uses of drugs or products in his presentation.

Climate Change May Result in More Lyme Disease

BY SHERRY BOSCHERT
EXPERT ANALYSIS FROM A DERMATOLOGY SEMINAR
LAS VEGAS – Climate change has expanded geo- graphic ranges of tick and parasite vectors, pushing into unfamiliar territory, Dr. Sigfrid A. Muller said at a seminar in Las Vegas and chair of the International Society of Dermatology.

Lyme disease has spread well into Canada, and leish- maniasis is moving north from Mexico into Texas, Ar- zona, Oklahoma, and Ohio. Reports of Chagas disease are increasing in the United States and Central and South America. Peru and Ecuador are seeing more Chag- rion’s disease, said Dr. Muller, a dermatologist in Las Vegas and chair of the International Society of Der-matology.

Global warming may be debated in the popular me- dia, but there is little controversy about it in the sci- entific literature, he said. June 2010 was the warmest month on record (combining global land and ocean av- erage surface temperatures) and the 304th consecutive month with a global temperature above the 20th-cen- tury average, according to the National Oceanic and At- mospheric Administration.

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