

20 **JUVENILE DERMATOMYOSITIS IS A DIFFERENT DISEASE IN CHILDREN UP TO 3 YEARS OF AGE AT ONSET THAN IN CHILDREN ABOVE 3 YEARS AT ONSET**

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Objective Juvenile dermatomyositis (JDM) is a different disease than dermatomyositis in adults in many ways including more vascular inflammation and vasculopathic thrombosis as previously described. The authors tested the hypothesis that

JDM disease course in children with disease onset at or below age 3 years may be different than that of children with disease onset at greater than 3 years of age.

Methods Institutional Review Board approval was obtained to retrospectively review the charts of 78 patients from age 0–18 years with JDM seen in pediatric rheumatology clinic at Nationwide Children's Hospital over the past 23 years the diagnosis was made by the treating pediatric rheumatologist. Not all the patients met the Bohan and Peter criteria, as muscle biopsy and EMG were not always performed. The data regarding disease course and outcome were collected as of the last clinic follow-up. Wilcoxon 2-sample test and χ^2 test and Fisher's exact test were used to compare continuous variables and categorical variables respectively.

Results The mean age of onset in the two groups was 27 months and 91 months. The mean times between onset of symptoms to diagnosis in younger and older age groups were 5.6 months and 4.5 months, respectively. The younger onset group had more females ($p=0.05$) and their disease onset occurred less frequently during the typical winter-spring seasons ($p=0.031$). The younger onset group was more likely to have a preceding fever ($p=0.029$) and family history of autoimmune diseases ($p=0.012$). The younger onset group was less likely to have heliotrope rash ($p=0.04$), Gottron's sign ($p=0.049$), capillary 2 loop abnormalities ($p=0.010$) or creatine kinase (CK, $p=0.022$), aspartate aminotransferase (AST, $p=0.021$) and aldolase ($p=0.035$) elevations. The younger children were more likely to have atypical histopathology ($p=0.02$) at presentation. The younger onset group was treated less often with pulse methylprednisolone at diagnosis ($p=0.043$) and less often with hydroxychloroquine ($p=0.035$). There were no differences between the two groups in the initial oral steroid dose ($p=0.8017$), number of patients who received methotrexate at diagnosis ($p=0.709$), and ever received other immunosuppressants ($p=0.323$). The mean ($p=0.06$) and maximum ($p=0.002$) duration of methotrexate therapy, and the mean ($p=0.06$) and maximum ($p=0.016$) duration of oral steroid therapy, was shorter in the younger group. Younger age group patients were more likely to experience a monocyclic course ($p=0.027$) and less likely to have active disease at 5 ($p=0.017$) and 10 years postdiagnosis ($p=0.019$). The younger patients were less likely to have osteonecrosis ($p=0.023$) and more likely to die of their disease.

Conclusion There were significant differences between JDM patients with disease onset at or below age 3 years, compared to their older counterparts. Younger patients in our cohort had fewer typical findings at diagnosis. They were more likely to experience a monocyclic course, a shorter and milder total disease course, and a shorter maximum duration of oral steroid and methotrexate therapy. Younger patients had predominantly monocyclic disease, and a lower proportion had active disease at 5 and 10 years. In spite of having milder disease course, patients with younger onset had a higher mortality and a similar complication rate compared to the older onset patients, except for osteonecrosis which was higher in the older onset group.