Skin involvement in juvenile dermatomyositis is associated with loss of end row nailfold capillary loops.

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OBJECTIVE: To determine associations of dermatological findings in children with juvenile dermatomyositis (JDM) with specific nailfold capillary (NFC) structural abnormalities.

METHODS: Sixty newly diagnosed, previously untreated children who met the Bohan-Peter criteria for definite JDM were seen between 1993 and 2002. They were classified by duration of untreated disease and by a disease activity score (DAS) composed of separate subscores for dermatological (DAS skin) and musculoskeletal (DAS muscle) findings. Routine NFC measurements yielded the number of end row loops, arboreal (bushy), and dilated capillary loops. Laboratory testing included muscle enzymes, von Willebrand Factor Antigen, and neopterin.

RESULTS: DAS skin, but not DAS muscle, was associated with NFC end row capillary loss ($r_s = -0.394$, $p = 0.008$). End row capillary loss (reflecting avascularity), arboreal (bushy), and dilated capillary loops (reflecting change in vascular morphology) were each associated with longer untreated symptom duration ($r_s = -0.401$, $r_s = 0.534$, $r_s = 0.371$).

CONCLUSION: End row capillary loss measured by NFC was associated with the dermatological, but not musculoskeletal manifestations of JDM, suggesting that damage to skin and muscle may each have distinct disease pathophysiology. In JDM, skin involvement indicates a vasculopathy that progresses with increasing duration of untreated disease and is not revealed by standard serological laboratory tests. We propose that the cutaneous manifestations of JDM are associated with vascular disease and warrant aggressive therapy.