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Original article

Nailfold capillary density is importantly associated over time with muscle and skin disease activity in juvenile dermatomyositis

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Abstract

Objectives. To investigate the longitudinal association of nailfold capillary density (NCD; as a potential marker of activity) with various other clinical measures of disease activity and to evaluate baseline NCD as a predictor of disease outcome in children with JDM.

Methods. Data from 809 clinic visits from 92 JDM patients were prospectively collected at each clinic visit over a time period of 5.5 years. The number of capillaries per millimetre at the distal nailfold was scored using a stereomicroscope. Disease activity was determined using the Childhood Myositis Assessment Scale (CMAS) and a modification of the validated disease activity score (DAS), which included three skin (SDAS) and three muscle (MDAS) criteria. An inception cohort subgroup ($n = 28$) with a baseline visit at diagnosis was analysed separately.

Results. Both DAS subscores, MDAS ($\beta = -0.04437$, $P < 0.0001$) and SDAS ($\beta = -0.1589$, $P < 0.0001$), as well as the CMAS ($\beta = 0.02165$, $P < 0.0001$) were significantly associated with loss of end row nailfold capillary over time (multiple regression mixed-model analysis). All patients in the inception subcohort showed a reduced baseline NCD (diagnostic sensitivity = 100%) that improved as the disease improved, but this did not predict longer term outcome or course of disease.

Conclusion. NCD is a marker of skin and muscle disease activity, and is an important measure of disease activity changes from visit to visit. Determination of capillary density may be useful when making treatment decisions. A decrease in NCD may be considered for inclusion in the diagnostic criteria due to its high sensitivity.

Key words: Juvenile dermatomyositis, Nailfold capillary density, Disease activity.

Introduction

JDM is a childhood idiopathic inflammatory autoimmune disease. Its estimated incidence is 2–3/million children/year [1, 2]. It is clinically characterized by progressive symmetrical proximal muscle weakness and a characteristic rash, but may also affect other organs [3–5].

Currently used diagnostic criteria are imperfect. The only published diagnostic criteria in current use are those defined by Bohan and Peter [6, 7] almost 30 years ago; these criteria require the presence of the pathognomonic skin rash and at least three other criteria (proximal muscle weakness, raised muscle enzymes, myopathic changes on the EMG or typical muscle biopsy changes).

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CLINICAL SCIENCE

However, a definitive diagnosis is not possible in all cases using these criteria. The heterogeneity of JDM means that not all affected individuals will display a characteristic skin rash, and some may have normal muscle strength and muscle enzymes despite evidence of myositis [4]. Muscle biopsy and even EMG are invasive investigations and are only moderately sensitive. In contemporary clinical practice, MRI is used by many clinicians as a non-invasive technique. Patients with no clinically apparent muscle signs may have changes on MRI, while not all subtle changes are recognized by MRI [4, 8–10]. New and better tools for diagnosis would seem to be of benefit.

Additionally, no single test is a reliable tool for monitoring disease activity. Two international collaborative networks, the PRINTO and the International Myositis Assessment and Clinical Studies Group (IMACS) have developed similar definitions of clinical improvement using a core set of variables to measure disease activity in JDM for use in therapeutic trials [11–13]. The PRINTO disease activity core set has been recently validated in a large number of JDM patients. Except for serum muscle enzymes, all candidate variables were found scientifically and clinically relevant with an overall good statistical performance [12]. These preliminary core sets of measures for disease activity will help standardize the assessment of patients with JDM and, in turn, improve the reliability of therapeutic and long-term outcome studies. However, both core sets still need further validation and each has few measures that can discriminate disease activity from damage. Patients who continue to have active disease are at risk of developing irreversible damage that may lead to permanent disability and impairment of quality of life. A reliable test for monitoring disease activity is important and would be useful when making treatment decisions.

The clinical features of JDM are largely due to a small-vessel vasculopathy, whereby activation and deposition of complement causes lysis of endomysial capillaries and perivascular inflammation; this is thought to lead sequentially to reduction of the capillary density with compensatory dilatation of the lumen of the remaining capillaries, muscle ischaemia, muscle fibre degeneration and muscle perifascicular atrophy [14].

It is possible that direct visualization of the capillaries may provide a sensitive diagnostic test as well as a valid indicator of disease activity [15–18]. Small blood vessels are most easily visualized at the nailfolds and around the teeth. Nailfold capillaroscopy is a non-invasive and inexpensive method to analyse microvascular abnormalities in autoimmune rheumatic diseases [19–22]. In JDM, nailfold capillaroscopy abnormalities have been proposed as a potential marker for more persistent and severe disease [23–28].

In our experience, nailfold capillary changes in JDM are almost universal, seem to change during the course of the disease and may reflect similar changes to those of the muscle capillaries. We wondered whether nailfold capillaroscopy could be used to reflect the degree of systemic blood vessel abnormalities [15, 24].

If nailfold capillary density (NCD) is shown to correlate highly with disease activity over time, capillary density may be helpful when making treatment decisions. In addition, capillaroscopy could be used to support a diagnosis of disease activity in patients who are otherwise difficult to clinically assess. If shown to be an accurate measure of disease activity, and a sensitive finding, a decrease in NCD may be helpful as a diagnostic criterion. Finally, capillary density may be a useful prognostic marker when studied early in the disease. The aim of our study was to investigate the correlation of NCD with various clinical measures of disease activity longitudinally, and to evaluate baseline NCD as a predictor of disease outcome.

Patients and methods

Population

We performed a retrospective cohort study of 92 patients (62 females and 30 males) followed in our subspecialty JDM clinic at The Hospital for Sick Children from July 2001 through September 2006 who met the Bohan and Peter classification criteria for definite or probable JDM [6, 7]. Children followed in the clinic with overlap syndrome ($n = 2$), MCTD ($n = 2$) and juvenile polymyositis ($n = 1$) were not included. The start date of July 2001 was chosen because at that time we obtained a new digital photographic microscope (see below) and we were able to record nailfold density at each clinic visit.

Patients were assessed and treated according to a defined protocol that has been in place since the inception of the clinic [29]. All clinic, nailfold capillaroscopy and laboratory data were prospectively collected and stored in a dedicated database.

New patients diagnosed during this time interval were considered to have had baseline nailfold capillaroscopy. They were analysed as an additional subsample (inception cohort) to evaluate whether baseline NCD can predict disease outcome. This study was approved by the Research Ethics Board at The Hospital for Sick Children.

Nailfold capillaroscopy

Before nailfold capillaroscopy each patient was acclimatized to room temperature (20–24°C) for at least 20 min. Nailfold capillaroscopy was carried out using a stereomicroscope (Nikon SMZ 800; Nikon, Mississauga, Canada) with colour video camera (Coolpix 990; Nikon) and television media attached. Wiping keratinized skin away from the distal nailfold with an alcohol swab prepped the right fourth digit (in our previous study, no significant difference in the NCD between various fingers in a subject had been found) [15]. If examination of the right fourth digit was technically impractical (e.g. intravenous in the right hand or injury), the left fourth digit was used. Immersion oil was applied to the nailfold just proximal to the nail for increasing transparency of the skin. NCD was determined by counting the total number of capillaries over the nailfold width and the mean capillary density per millimetre was

determined. A capillary density of <6 capillary loops/mm was considered abnormal [22]. In all patients, nailfold capillaroscopy was performed and analysed by a trained research assistant, who did not have knowledge of the disease activity status of the patients.

The changes seen in JDM at the nailfold include capillary dropout, branching and dilatation, areas of haemorrhage and decrease in the number of vessels per millimetre [19, 24, 26]. Because capillary density is the only good objective measure of nailfold vessel pathology, which is not significantly affected by age and gender, it was chosen for judgement of severity and gradation of nailfold vasculopathy [23, 30].

Assessment and outcome parameters

Disease activity

At each visit, patients had a standardized assessment of skin, muscle and global disease activity. Disease activity was determined using the manual muscle test (MMT), the Childhood Myositis Assessment Scale (CMAS) and levels of muscle enzymes [creatine kinase (CK), alanine aminotransferase (ALT) and aspartate aminotransferase (AST)] [13, 31–33]. In addition, we assigned a modification of the validated disease activity score (DAS) that included clinical parameters for three dermatological (SDAS) and three musculoskeletal (MDAS) criteria [34]. Zero to 4 points could be given on the basis of dermatological findings and 0–7 points on the basis of musculoskeletal findings, whereby a higher score is associated with a higher disease activity (Table 1). This modified DAS, which we have previously used in a study, is one that can be applied in chart and database review [35].

Quality of life

To evaluate quality of life and general function, the Childhood HAQ (CHAQ) and its visual analogue scales for disease activity, pain and disease impact as well as visual analogue scales for global quality of life and health-related Quality of My Life (QoML questionnaire) were recorded by the patient/parent at each visit [36–39].

Statistical analysis

The data were first explored using longitudinal plots and smoothers, histograms, data summaries such as medians, ranges, means and S.D. A linear mixed regression model was then developed for each outcome, using the mixed procedure in statistical analysis system (SAS) software, version 9.1 (SAS, Cary, NC, USA). This kind of model incorporates the within-subject correlation structure that is an inherent characteristic of longitudinal data. The main predictor in each model was NCD—time was also included as an independent variable—as a linear or quadratic effect, depending on the case. Interactions between time and capillary density were also explored in each model.

Residual analysis was used to validate the models. Where the assumptions were not verified, log or square root transformations were used on the outcomes. If these failed to normalize the residuals, a general linear mixed model was fitted using the Glimmix procedure in SAS.

In those cases where all of the above failed to provide a valid model, the outcome was dichotomized and a generalized estimating equation (GEE) model was fitted using the Genmod procedure in SAS. This model is also designed to take into account the within-subject correlation structure of the outcome.

In a separate set of models, the impact of baseline capillary density on important longer term outcomes was explored similarly. The Cox proportional hazards regression model was used to determine whether baseline capillary density is a predictor of time to clinical quiescence and remission.

TABLE 1 Modified DAS

MDAS	Score	SDAS	Score
Muscle strength (maximum 3 points)		Erythema (maximum 2 points)	
No muscle weakness (gMMT 68–70)	0	No erythema	0
Minimal muscle weakness (gMMT 63–67)	1	Local erythema (joints and face)	1
Moderate muscle weakness (gMMT 56–62)	2	Widespread erythema	2
Severe muscle weakness (gMMT 0–55)	3		
Functional status (maximum 3 points)		Heliotrope rash (maximum 1 point)	
No limitations on activity	0	Absent	0
Activities limited at the extra-curricular level	1	Present	1
Activities limited at the school/work level	2		
Activities limited at the self-care level	3		
Arthritis (maximum 1 point)		Gottron's papules (maximum 1 point)	
Absent	0	Absent	0
Present	1	Present	1
MDAS: 0–7 points		SDAS: 0–4 points	
Total DAS: 0–11 points			

The modified validated DASs include clinical parameters for three dermatological (SDAS) and three musculoskeletal (MDAS) criteria [34, 35]. gMMT is the manual muscle testing score [45].

Results

Patient characteristics

The cohort consisted of 92 patients with definite or probable JDM. The majority of patients with probable JDM had findings consistent with myositis on MRI (13/13 tested).

There was a total of 809 clinic visits with concurrent nailfold capillaroscopy collected over a time period of 5.5 years. The number of follow-up visits per patient ranged from 1 to 26 with a mean/median of 8.8/9.0. The time between diagnosis and the first-recorded NCD was between −1.4 and 176.4 months (median 18.6 months, mean 40.1 months). At the first testing of the whole cohort, the mean/median NCD was 5.4/5.5/mm with a range of 0.7–10.6/mm. The data were mostly complete for the disease activity instruments: SDAS 99.1% (802 visits); MDAS 93.8% (759 visits); DAS 93.2% (754 visits) and CMAS 83.4% (675 visits). Quality of life and general physical function measurements were available for ~75% of the visits. Demographics, diagnostic tests and disease activity parameters of the whole cohort at first nailfold examination are summarized in Table 2.

Correlation of disease activity and NCD over time

Longitudinal data analysis with time as a continuous measure (using all visits) showed that the loss of end row nailfold capillary was moderately associated with skin disease activity (SDAS: $\beta = -0.1589$, $P < 0.0001$) and more modestly associated with musculoskeletal involvement/function (MDAS: $\beta = -0.04437$, $P < 0.0001$; CMAS: $\beta = 0.02165$, $P < 0.0001$) (Fig. 1A, B and C). This relationship was slightly more pronounced at the beginning of the disease course and relatively consistent over time (Fig. 2).

Some outcomes were highly skewed and were transformed to binary variables for this analysis. For example, health-related quality of life was transformed to a binary variable—normal/abnormal. For each capillary per millimetre increase in nailfold density the likelihood of having normal health-related quality of life increased by 24% [odds ratio (OR) = 1.24, $P = 0.0009$]. Likewise, overall quality of life (OR for normal = 1.24, $P = 0.003$), CHAQ (OR for disabled = 0.86, $P = 0.03$), MMT (OR for normal = 1.29, $P = 0.0004$), overall disease severity (OR for abnormal = 0.79, $P = 0.0004$) and pain (OR for abnormal = 0.82, $P = 0.001$) were related to NCD.

Nailfold capillaroscopy in the inception cohort

During the time interval 28 children were newly diagnosed and had baseline nailfold capillaroscopy. All of these patients showed a reduced baseline NCD (diagnostic sensitivity = 1.0) that ranged from 0.7 to 5.6 (mean 3.4, median 3.1). Patient characteristics are summarized in Table 3.

Treatment effect on NCD

The inception cohort had capillary density measurements before and after treatment. With treatment, children showed a notable improvement in baseline NCD over time (reflecting sensitivity to change of the nailfold measurements) as seen by a moderate to large effect size of 0.69 [standardized response mean (SRM) = 0.53] after 3 months, 1.27 (SRM = 1.04) after 6 months, 1.32 (SRM = 0.98) after 9 months and 1.57 (SRM = 1.21) after 12 months. Six of 22 children who had a measurement (27.3%) normalized the NCD ($\geqslant$6 capillaries/mm) after 6 months and 4 of 17 children (a further 23.5%) after 12 months on treatment (Fig. 3).

Nailfold capillaroscopy as predictor of disease outcome

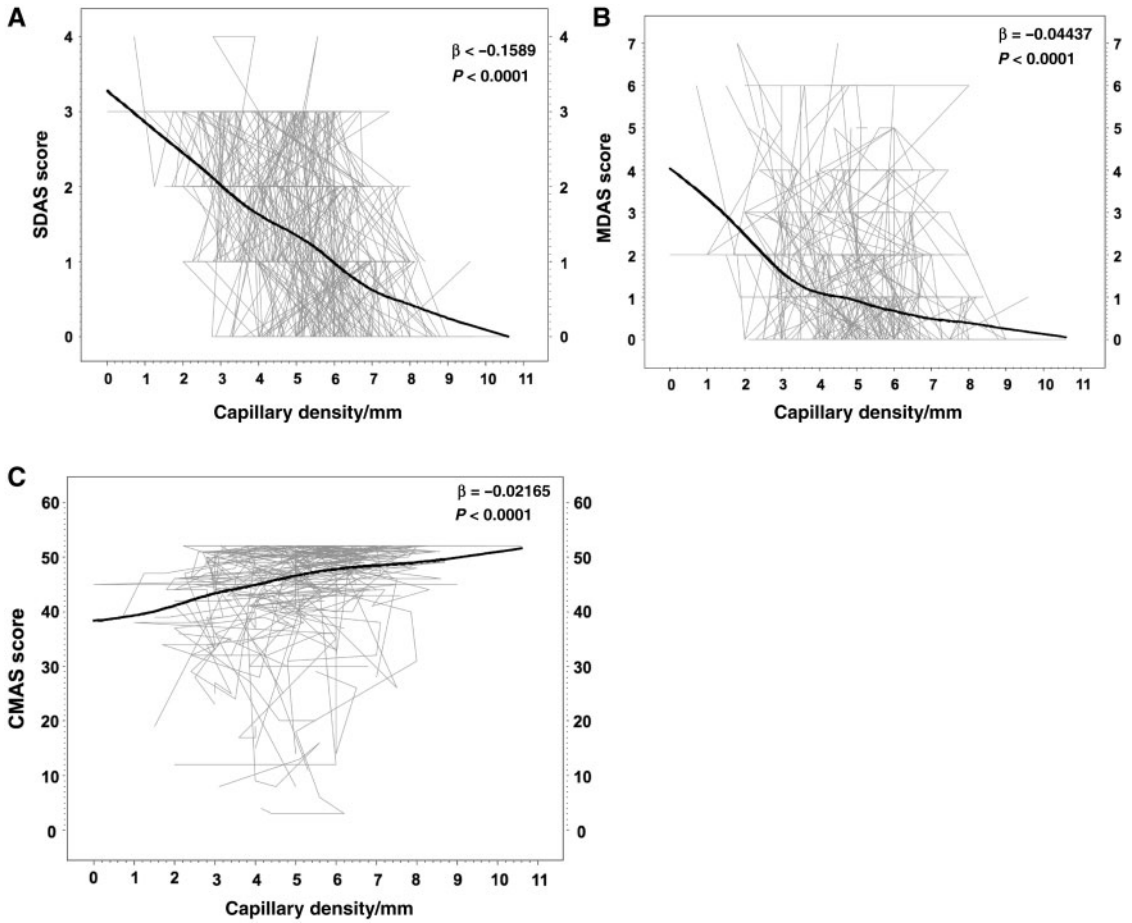
In our inception cohort, we were unable to demonstrate that NCD predicts longer term outcome or course of

Table 2 Demographics, diagnostic tests and disease activity parameter at first nailfold examination in 92 patients with JDM

Age at diagnosis, mean/median, years	7.6/7.1
Number of females/number of males	62/30
Definite JDM, *n* (%)	77 (83.7)[a]
Probable JDM, *n* (%)	15 (16.3)
Gottron's papules and/or heliotrope rash	91/92 (98.9)[a]
Muscle weakness	90/91 (98.9)
Muscle enzyme elevation	85/92 (92.4)
Abnormal EMG	64/76 (84.2)
Abnormal muscle biopsy	44/46 (95.7)
Abnormal MRI	67/71 (94.4)
Disease activity parameter at first nailfold examination, mean/median (range)	
MDAS (0–7) (n = 82)	1.5/0 (0–6)
SDAS (0–4) (n = 91)	1.5/2.0 (0–4)
DAS (0–11) (n = 82)	3.0/2.0 (0–10)
CMAS (0–52) (n = 66)	40.5/46 (4–52)
CHAQ (0–3) (n = 75)	0.36/0 (0–2.6)

[a]One patient did not present with a characteristic rash; however, the muscle biopsy showed typical findings of DM. Therefore, the patient was classified as definite JDM.

FIG. 1 Correlation between SDAS, MDAS, CMAS and capillary density. (**A**) Correlation between SDAS and capillary density. The *y*-axis shows the SDAS (no disease activity = 0, severe disease activity = 4), the *x*-axis shows the capillary density from very abnormal to normal (normal 6–11). Longitudinal data analysis with time as a continuous measure (using all visits) showed that the loss of end row nailfold capillary was moderately associated with SDAS. (**B**) Correlation between MDAS and capillary density. The *y*-axis shows the MDAS (no disease activity = 0, severe disease activity = 7), the *x*-axis shows the capillary density from very abnormal to normal (normal 6–11). Longitudinal data analysis with time as a continuous measure (using all visits) showed that the loss of end row nailfold capillary was more modestly associated with musculoskeletal activity. (**C**) Correlation between CMAS and capillary density. The *y*-axis shows the CMAS, in contrast to the SDAS and MDAS, going from very abnormal to normal muscle function (normal muscle function 50–52), and the *x*-axis shows the capillary density from very abnormal to normal (normal 6–11). Longitudinal data analysis with time as a continuous measure (using all visits) showed that the loss of end row nailfold capillary was more modestly associated with muscle function. Grey lines represent individual subjects; the bold line is the overall fitted line from the longitudinal model.



disease. During the observation time 8 of 28 patients developed normal muscle strength, muscle enzymes and skin (clinical quiescence). Three of 28 patients went into remission off medication. Remission was defined as a clinical state in which rash (heliotrope rash, Gottron's papules or skin ulcers) is absent, there is no evidence of active myositis (normal strength and normal muscle enzyme levels) or arthritis and which is maintained in the absence of all immunosuppressive medications for a minimum of 6 months [40]. We were unable to demonstrate that baseline NCD predicts time to clinical quiescence [risk ratio/hazard ratio (HR) = 0.93; 95% CI 0.54, 1.57; $P = 0.80$] or time to remission (HR = 1.2; 95% CI 0.49, 2.87; $P = 0.67$).

Discussion

We have found that, for JDM patients, NCD is a good indication of skin disease activity as well as muscle disease activity. This relationship appears to be relatively consistent over time. In addition, all of our newly diagnosed patients had a decreased baseline NCD that improved notably with treatment. Therefore, reduced NCD is a highly sensitive diagnostic marker for JDM, and capillary

microscopy is a responsive measure of disease activity over time. Perhaps because of the smaller subsample used and a relatively short follow-up, we were unable to demonstrate an expected ability of NCD to predict outcome, disease course or time to remission.

These results are consistent with our previous finding of correlations between NCD and both skin and muscle disease activity in a cross-sectional study of 42 children with juvenile idiopathic inflammatory myopathies (38 with typical JDM) from two centres [15].

Based on these data, nailfold capillary changes likely reflect the degree of systemic blood vessel abnormality in JDM. NCD seems to be an accurate objective measure of disease activity and a non-invasive reliable test for

Fig. 2 Loess smoothing of the relationship between total DAS and capillary density is slightly more pronounced at the beginning of the disease course and relatively consistent over time. The *y*-axis shows overall disease activity (worsening along the indicated arrow); the *x*-axis shows time (progressing along the indicated arrow); the *z*-axis shows capillary density (increasing, i.e. becoming more normal, along the indicated arrow).

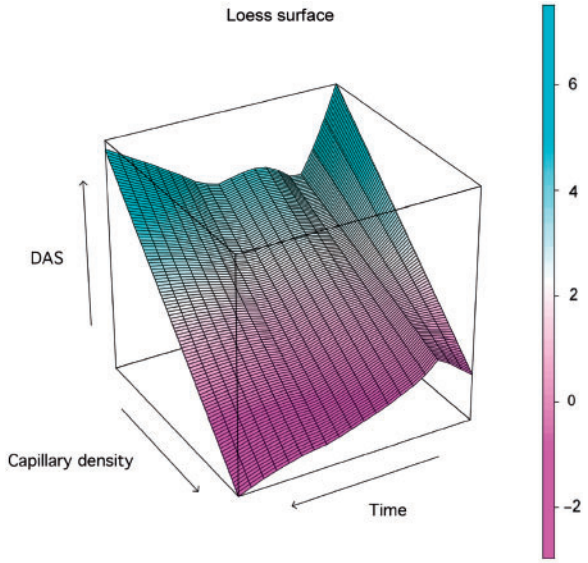


Fig. 3 Inception cohort—improvement of baseline capillary density over time. This is a box-and-whisker plot. The whiskers show the limits of the data while the shoulders of the boxes show the 25th and 50th percentile values for the cohort. The central (bold) line of the box indicates the median value. This graph shows the same children followed from visit to visit over 12 months. The *y*-axis shows the capillary density from very abnormal to normal (normal 6–11), and the *x*-axis shows time from baseline, and Months 3, 6, 9 and 12. With treatment, children showed considerable improvement in baseline NCD over time as seen by a moderate to large effect size (ES).

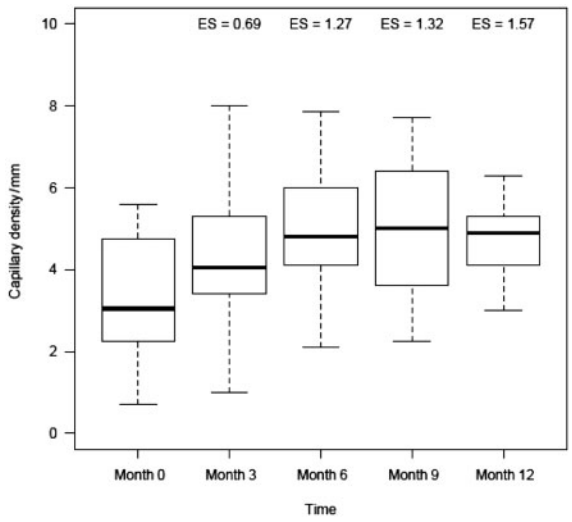


Table 3 Demographics, disease activity at the first visit and treatment of the inception cohort ($n = 28$)

Age at diagnosis, mean/median, years	9.9/10.3
Number of females/number of males	20/8
Definite JDM, *n* (%)	20 (71.4)
Probable JDM, *n* (%)	8 (28.6)
Duration of follow-up, mean/median (range), months	23.8/21.3 (1.4–54.3)
Number of clinical visits per patient, mean/median (range) (total 282)	10.1/9.0 (3–26)
NCD, mean/median (range)	3.4/3.1 (0.7–5.6)
Disease activity parameter at the first visit, mean/median (range)	
MDAS (0–7) ($n = 22$)	4.1/4.5 (0–6)
SDAS (0–4) ($n = 28$)	2.4/3.0 (0–4)
DAS (0–11) ($n = 22$)	6.5/7.0 (1–10)
CMAS (0–52) ($n = 26$)	29.1/30.5 (4–52)
CHAQ (0–3) ($n = 20$)	1.0/0.9 (0–2.6)
Treatment, *n* (%)	
Corticosteroids	28 (100)
MTX	28 (100)
IVIG	8 (28.6)
Other	2 (5.6)

monitoring disease activity. Therefore, determination of NCD has important clinical implications: it provides clinicians with another sensitive parameter of disease activity; it may be used to support a diagnosis of disease activity in patients who are otherwise difficult to assess; and it can be used as a diagnostic criterion given its very high sensitivity.

Several studies have shown that a delay of treatment, or the administration of low-dose therapy, predicts poor functional outcome and the development of calcinosis [41–43]. Persistent nailfold changes may be a good marker of a persistent active vasculopathy and identify patients at risk of developing irreversible damage and long-term complications if left untreated. Since it is a marker of disease activity, persistent decreased NCD might indicate therapy, even in the absence of obvious clinical signs like muscle weakness or JDM rash.

The relationship between nailfold capillary abnormalities and muscle vasculopathy has been demonstrated by Spencer *et al*. [17, 18]. In contrast, Pachman's group was unable to confirm these findings. They found a strong association with the cutaneous but not with the musculoskeletal signs of JDM [27, 28] . For the determination of skin and muscle disease activity that group used a clinically based DAS [34]. Nine points are given on the basis of clinical dermatological findings and 11 points based on musculoskeletal findings. The musculoskeletal components include areas of weakness (points are given per weakness area noted) but not for grade of the muscle strength. Perhaps the discrepant findings regarding the relationship between capillaroscopy and muscle activity are related to the different ways of measuring muscle activity; our modified MDAS includes a gradation of muscle weakness.

All of our inception subsample had a decreased baseline NCD. Results reported by Scheja *et al*. [23] are consistent with our findings. The group compared the nailfold density in JDM patients, children with MCTD and healthy controls. In that study, the disease duration of JDM was 1–4 years (median 1 year). All seven children with JDM had a significantly reduced NCD (median 2.5, range 1.4–4.3 loops/mm) when compared with healthy controls (median 6.8, range 5.3–8.0 loops/mm) [23]. These results indicate that nailfold abnormalities occur early in JDM and are almost universal; capillary microscopy therefore has diagnostic utility, particularly in patients who are otherwise difficult to assess. Importantly, because very sensitive tests can rule out conditions if the test is negative, the very high sensitivity of nailfold abnormalities makes a diagnosis of JDM less likely if nailfold capillaroscopy reveals normal density—although Ostrowski *et al.* [44] found that if patients have a short duration of symptoms they may still have normal NCD.

Our clinical observation is that improvement in skin rash and weakness may change more quickly than nailfold capillary counts. It is likely that improved strength and rash reflect improvement in a number of pathological mechanisms, and not just improved perfusion related to better capillary density; capillary density should not be the only measure used to determine disease activity. Nevertheless, in our inception cohort subsample, we found capillary density to be a very responsive measure of treatment response. Moreover, the relationship between capillary density and disease activity seems to be relatively stable over time.

We were unable to demonstrate that NCD predicts outcome, disease course or time to remission. This may be because of the smaller subsample used and short length of follow-up. During the observation time only 8 of 28 patients developed clinical quiescence and only 3 patients went into remission. In another study, at 6 months, abnormal nailfold capillaries in addition to Gottron's papules predicted a longer time to remission [40]. In another cohort, nailfold capillaroscopy could not predict the course of disease or time to remission, but a longer duration of untreated disease was found to be associated with persistent nailfold capillaroscopy abnormalities and with a non-unicyclic disease course [28]. This report and the results of our studies perhaps suggest that, rather than the severity of nailfold capillary abnormalities at disease onset, it is the persistence that has important prognostic value.

Our findings must be interpreted in the context of potential limitations of our study. Many subjects had their first nailfold capillaroscopy measured late in their disease course and had inactive disease at the time of that first examination. This may perhaps explain why we did not find a stronger association between nailfold capillary abnormalities and quality of life assessments (e.g. more than half the subjects had a normal CHAQ at first nailfold assessment). The patient group with baseline nailfold capillaroscopy was small with a relatively short time of follow-up; we were unable to demonstrate that capillaroscopy at diagnosis was of predictive value, but this may reflect a type II error. We did not attempt to correlate features of muscle biopsy or MRI with nailfold capillaroscopy—findings that might have provided us with additional information about the reasons for the associations that we saw.

In conclusion, NCD is importantly related to skin activity as well as to muscle activity and is a valid and responsive measure of disease activity changes from visit to visit. Nailfold capillary abnormalities are a sensitive diagnostic marker for JDM. Based on our findings, we believe that nailfold capillary changes reflect the degree of systemic blood vessel abnormality in JDM. Nailfold capillaroscopy is an easy-to-perform, non-invasive technique providing a sensitive diagnostic test as well as a valid indicator of disease activity and may be useful when making treatment decisions. Given the high sensitivity, a decrease in NCD may be considered for inclusion in the diagnostic criteria.

Rheumatology key messages

- In JDM, reduced NCD is a sensitive diagnostic marker.
- NCD correlates strongly with disease activity, having an important impact in making treatment decisions.

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