Skin Capillary Abnormalities As Indicators of Organ Involvement in Scleroderma (Systemic Sclerosis), Raynaud's Syndrome and Dermatomyositis

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Forty-four study patients with scleroderma (systemic sclerosis) (28 patients), Raynaud's syndrome (13 patients) or dermatomyositis (three patients) were observed for skin capillary abnormalities by widefield microscopy and compared with three control groups of 20 subjects each: (1) patients with other rheumatic disease, (2) hospitalized patients with nonrheumatic conditions, and (3) healthy volunteers. The distinctive microvascular pattern (dilated and distorted capillary loops alternating with avascular areas) previously reported in scleroderma and dermatomyositis was observed almost exclusively in the study patients. The severity of capillary abnormalities varied among the diagnostic subgroups, and a positive correlation was found between the degree and extent of abnormal microvascular patterns and multisystem involvement.

On this basis, widefield nailfold capillary observations are proposed as a simple, inexpensive, reproducible technic for making an improved early diagnosis and predicting multisystem involvement in scleroderma, Raynaud's syndrome and dermatomyositis, presently a group of loosely associated and overlapping connective tissue disorders which often defy early and precise diagnosis.

The diagnosis of rheumatic disorders has been improved by the introduction of the criteria for acute rheumatic fever, rheumatoid arthritis, systemic lupus erythematosus, gout and pseudogout. Such criteria are lacking for several loosely associated and often overlapping syndromes: scleroderma (systemic sclerosis), Raynaud's syndrome without a definable connective tissue disorder and dermatomyositis.

Raynaud's phenomenon occurs in approximately 80 to 90 per cent of the patients with scleroderma [1] and often precedes other symptoms and signs. At an early stage it is, therefore, difficult to distinguish between patients with so-called "Raynaud's disease" (pathology limited to extremities) and those in whom scleroderma later develops (a multisystem disorder whose more serious prognosis depends on visceral involvement) [2–5]. A common denominator of all target organ involvement in scleroderma appears at the microcirculatory level and consists of distinctive lesions of small arteries and arterioles [1,6,7].

As a potential diagnostic aid, the morphology of skin and other surface capillaries has been known to be abnormal in rheumatic diseases for a half century [8–10]. The favorite site of observation has been the nailfold capillary bed. Similar abnormalities of nailfold capillaries in scleroderma, Raynaud's syndrome and dermatomyositis have

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been reported by a number of investigators [8–18]; principal findings have consisted of marked dilatation of certain capillary loops. Attempts to distinguish between Raynaud's "disease" and scleroderma on the basis of capillary findings have not been conclusive, and the capillary abnormalities have not before been correlated with organ involvement.

By using widefield examination and evaluating patterns of microvascular abnormalities in several areas of finger skin rather than elementary capillary lesions in a limited area of nailfold, a characteristic and similar pattern of capillary abnormalities has been observed in scleroderma, dermatomyositis and Raynaud's syndrome which is distinct from patterns seen in systemic lupus erythematosus and rheumatoid arthritis [19–21]. The connection between small artery and arteriolar lesions observed histopathologically in internal organs, and the capillary abnormalities seen with in vivo capillaroscopic methods in the skin is not known. A correlation between visceral involvement in scleroderma-Raynaud's syndrome-dermatomyositis and the extent of finger capillary abnormalities would establish a link between the arterial and capillary abnormalities and suggest a common or related pathogenesis.

For these reasons the widefield nailfold capillary patterns of all patients with scleroderma-Raynaud's syndrome-dermatomyositis referred to a rheumatic disease clinic were studied and compared with three selected control groups to determine (1) the specificity of the abnormal capillary pattern for scleroderma-Raynaud's syndrome-dermatomyositis and (2) the relationship of the severity of capillary abnormalities to the severity of the clinical syndrome. The distinctive capillary pattern was found to be specific for scleroderma-Raynaud's syndrome-dermatomyositis and a positive correlation was found between the severity of the capillary changes and the clinical syndrome, as determined by the number of discrete organ systems involved.

PATIENTS AND METHODS

Study Patients. The study group consisted of all patients seen in the Edward Daniels Faulkner Arthritis Clinic during the 10 month period between July 1, 1973, and May 1, 1974, in whom the diagnosis of scleroderma, Raynaud's syndrome or dermatomyositis was either definite or tentative. A total of 50 patients was seen; of these, six were excluded (three with scleroderma, two with dermatomyositis, one with Raynaud's syndrome) because it was not possible to complete all examinations required. In the 44 study patients in whom complete and often repeated skin capillary observations were made, the eventual diagnosis was scleroderma in 28, Raynaud's syndrome in 13 and dermatomyositis in three. Subjects ranged in age from 20 to 71 years and consisted of 40 women (mean age 44.9 years) and four men (mean age 39.8 years).

The major criterion for the diagnosis of scleroderma was taut skin involving diffusely the fingers, hands, arms and, in some patients, the face and trunk as well.

Twenty-five of the 28 patients with scleroderma had diffuse scleroderma of the extremities and trunk. Three of the 28 had taut skin distal to the metacarpophalangeal joints only (sclerodactyly) and, on this basis, could qualify for the designation CREST syndrome (calcinosis, Raynaud's phenomenon, esophageal hypomotility, sclerodactyly and telangiectasia [22,23]). In all three of these patients, two organ systems other than skin were involved.

The diagnosis of Raynaud's syndrome was based on the observation by a physician of the characteristic biphasic vasomotor response (pallor or cyanosis during vasoconstriction and suffusion, erythema or edema during reactive vasodilatation) in a patient in whom clinical and laboratory

TABLE I Study Group: Clinical Criteria

System	Criteria of Involvement*
Peripheral vascular	1. Presence of Raynaud's phenomenon documented by a physician other than capillary observer
Cutaneous	1. Typical symmetrical skin changes (hidebound dermis, atrophic epidermis with loss of appendages) 2. Sclerodactyly on physical examination by physician observer 3. Skin biopsy specimen consistent with scleroderma
Gastrointestinal	1. Decreased esophageal motility on x-ray series of upper gastrointestinal tract 2. Widened duodenum on x-ray series of upper gastrointestinal tract 3. Sacculations of colon on roentgenogram of lower gastrointestinal tract
Pulmonary	1. Bibasilar pulmonary markings on the roentgenogram of chest with no other underlying cause 2. Abnormal diffusion capacity on pulmonary function tests 3. Vital capacity <70 per cent predicted
Cardiac	1. Conduction defect on electrocardiogram 2. Low voltage throughout on electrocardiogram 3. Pericarditis 4. Cardiomegaly
Muscular	1. Proximal muscle weakness by physician observer 2. Creatine phosphokinase activity >50 IU/liter 3. Electromyogram consistent with myositis 4. Myositis on muscle biopsy
Renal	1. Recent onset (<6 mo) of increased blood pressure (diastolic >90 mm Hg) 2. Elevated blood urea nitrogen and creatinine levels on >1 determination 3. Abnormal urinalysis (>1+ proteinuria, >2 red blood cells, casts in sediment)

*A single criterion was considered positive in all systems except renal, in which two criteria were required.

evaluation did not support the diagnosis of rheumatoid arthritis, systemic lupus erythematosus, scleroderma or dermatomyositis, and in whom intravascular (cryoglobulin, cold agglutinin), mechanical (machine operators) and toxic (vinyl chloride) causes were not present.

The diagnosis of dermatomyositis was based on the presence of characteristic skin (dermatitis of face, trunk or hands and/or subcutaneous calcification) and muscle (criteria for myositis as specified in Table I) involvement. Although dermatitis was essential for the diagnosis of dermatomyositis, it was not included as an independent organ involvement in the data presented.

Patients in the study group were receiving a variety of symptomatic therapies. The wide variety prevented a systematic analysis of therapy. Previous observations have not suggested any specific therapy that might be correlated with capillary abnormalities of the type seen in scleroderma-Raynaud's syndrome-dermatomyositis.

Control Groups. Three separate groups of controls, each consisting of 20 subjects, were observed.

Group 1: Rheumatic controls consisted of five men (mean age 51.5 years) and 15 women (mean age 42.9 years) from the clinic, with the diagnosis of rheumatoid arthritis in nine patients, psoriatic arthritis in two patients, systemic lupus erythematosus in seven patients, Sjögren's syndrome in one patient and periarteritis nodosa in one patient. Three of the patients with systemic lupus erythematosus had skin involvement. No patient in this group had Raynaud's syndrome or sclerodactyly.

Group 2: Hospitalized control subjects consisted of 20 women from 17 to 77 years of age (mean age 46.7 years) who were hospitalized at the Columbia-Presbyterian Medical Center for other than rheumatic disease and who gave no history of inflammatory arthritis. Six were recovering from uneventful obstetrical deliveries; the remaining 14 were hospitalized for one of the following diseases: fever of unknown origin, cystitis, reticulum cell sarcoma, Hodgkin's disease, bullous pemphigoid, cancer of the breast, osteoarthritis of the spine, pyelonephritis, giardiasis, cancer of the stomach, peripheral neuropathy, macroglobulinemia, cerebellar ataxia and cerebral vascular accident.

Group 3: Healthy control subjects consisted of three men (mean age 36.3 years) and 17 women (mean age 43.5 years) who were nonhospitalized volunteers with no history of rheumatic complaints, dysphagia or Raynaud's phenomenon. Black patients were not studied because of the inability to visualize capillaries through deeply pigmented skin.

Technics. Each patient was examined by inspection for visible abnormalities of the hands and by widefield microscopy (12X) at six standard sites on each hand including the nailfold and the remainder of the dorsum of the distal phalanx, and the finger pad of the fourth finger; the nailfold-distal

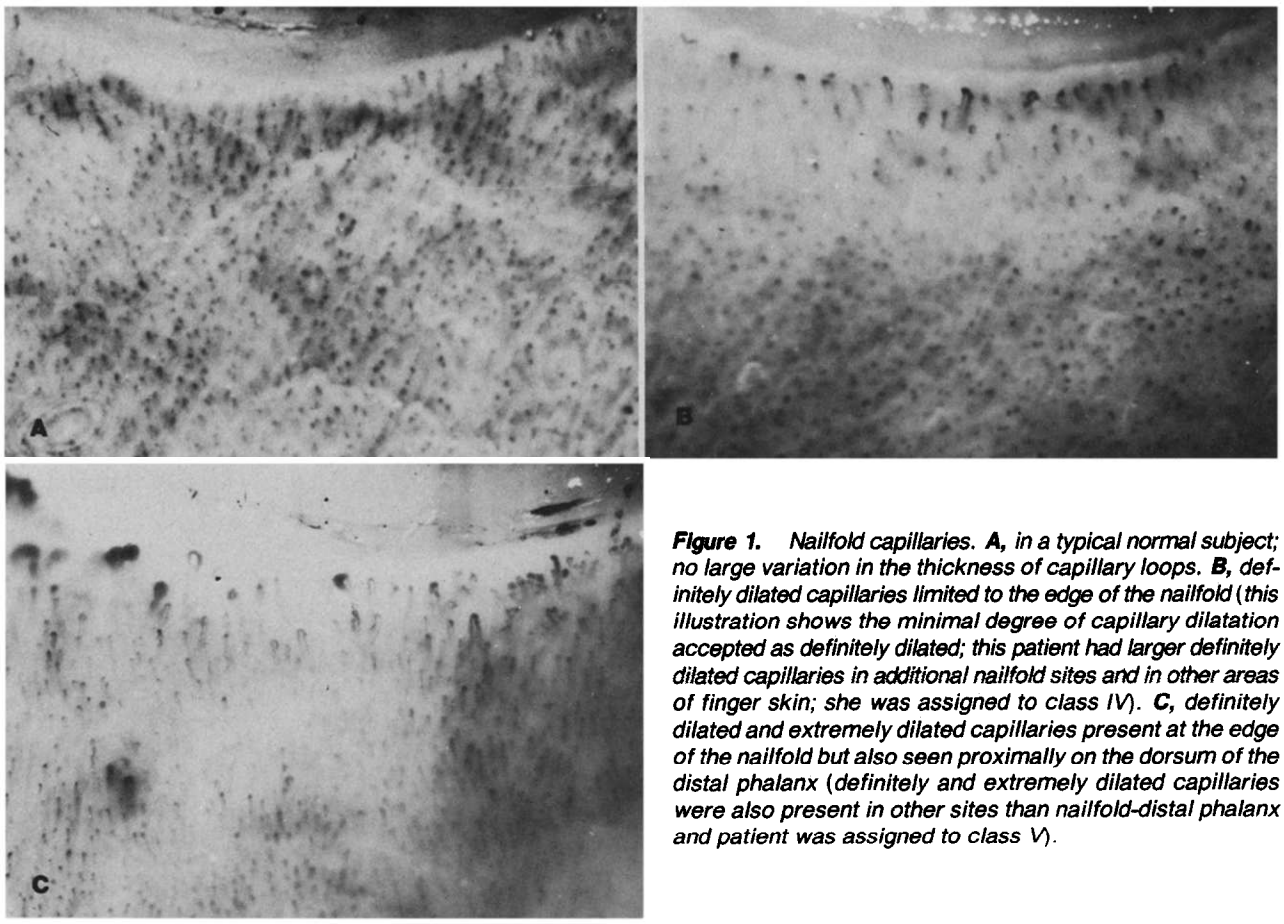

Figure 1. *Nailfold capillaries.* ***A,*** *in a typical normal subject; no large variation in the thickness of capillary loops.* ***B,*** *definitely dilated capillaries limited to the edge of the nailfold (this illustration shows the minimal degree of capillary dilatation accepted as definitely dilated; this patient had larger definitely dilated capillaries in additional nailfold sites and in other areas of finger skin; she was assigned to class IV).* ***C,*** *definitely dilated and extremely dilated capillaries present at the edge of the nailfold but also seen proximally on the dorsum of the distal phalanx (definitely and extremely dilated capillaries were also present in other sites than nailfold-distal phalanx and patient was assigned to class V).*

phalanx, the dorsum of the middle phalanx and the dorsal aspect of the middle joint of the third finger; and an area of dorsal skin 1.5 cm proximal to the fourth metacarpalphalangeal joint. Immersion oil was applied to each site prior to microscopic examination to aid in visualization of the vessels. A photographic record was made of each site examined on the left hand, using high speed Ektachrome film, a single lens reflex camera with macro lens and bellows, and a low voltage, low heat light source (to reduce local heat-induced vasodilatation) so that the results could be evaluated by a trained observer unaware of the patients' clinical status. Final magnification was between 3X and 4X on the negative. Technics were similar to those previously reported [19,20,24]. Four patients could not be fully photographed due to hand deformities secondary to scleroderma.

Observations made at each site examined included the presence or absence of dilated capillary loops, visible subpapillary plexus, relative loss of capillaries and dilated venules. These changes have previously been described in connective tissue disease [19,20]. Capillary loop size was designated normal, slightly dilated, definitely dilated (loop increased in size but loop form preserved), and extremely dilated (capillaries increased several times in size and capillary loops distorted). In addition, any areas that appeared visibly abnormal were observed microscopically; if abnormalities were seen, photographs were taken.

At the time of capillary examination, the following data were obtained from each patient: history and duration of Raynaud's phenomenon, stiffness and/or swelling of finger and wrist joints, muscle aches and weakness, dysphagia and dyspnea on exertion.

Clinical and Laboratory Evaluation. The following laboratory data, obtained from patients in the study group during the two months prior to study, were compiled: roentgenogram of the chest, pulmonary function studies, electrocardiogram, barium contrast roentgenographic study of upper and lower gastrointestinal tract, urinalysis, blood pressure, serum creatinine and blood urea nitrogen. Results of creatine phosphokinase activity, electromyogram and skin biopsy specimens obtained within the year before the examination were evaluated. Bibasilar pulmonary markings, decreased esophageal motility or widened duodenum, or colonic sacculations on roentgenographic examination at any previous time were considered positive.

Seven systems were evaluated in study patients from the clinical data. These included peripheral vascular, cutaneous, gastrointestinal, pulmonary, cardiac, muscular and renal, which were determined to be involved by the criteria listed in Table I. These criteria are similar in all respects to those proposed by Medsger et al. [4]. Data were adequate to determine system noninvolvement in 88 per cent; the remaining 12 per cent with missing data were randomly distributed with

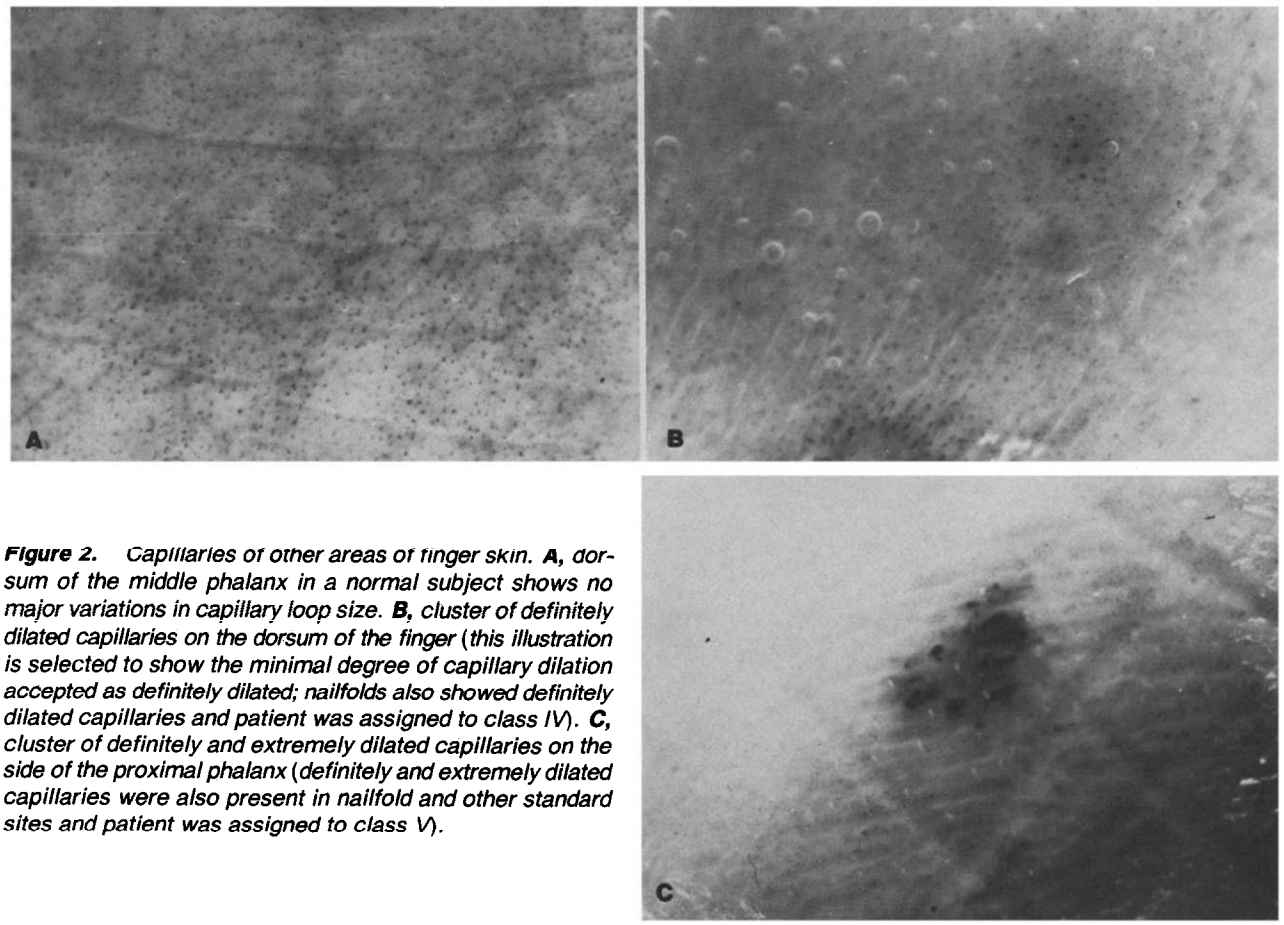

Figure 2. *Capillaries of other areas of finger skin.* ***A,*** *dorsum of the middle phalanx in a normal subject shows no major variations in capillary loop size.* ***B,*** *cluster of definitely dilated capillaries on the dorsum of the finger (this illustration is selected to show the minimal degree of capillary dilation accepted as definitely dilated; nailfolds also showed definitely dilated capillaries and patient was assigned to class IV).* ***C,*** *cluster of definitely and extremely dilated capillaries on the side of the proximal phalanx (definitely and extremely dilated capillaries were also present in nailfold and other standard sites and patient was assigned to class V).*

TABLE II Clinical Features and Capillary Class in the Study Group

Diagnosis	Total No. of Patients	Capillary Class	No. of Patients	Organs Involved		V*	Duration (yr)	
				Mean	Range		Mean	Range
Raynaud's syndrome	13	I	6	1	1	0	7	<1 – 10
		II	6	2	1 – 3	3	8	<1 – 25
		IV	1	. . .	3	1	<1	. . .
Scleroderma	28	I	2†	. . .	2 and 5	1	<1	. . .
		II	1	. . .	2	0	>20	. . .
		III	3	4	3 – 4	3	. . .	<1 – 25
		IV	7	4	2 – 5	6	5	2 – 13
		V	15‡	4	3 – 7	15	9	<1 – 20
Dermatomyositis	3	III	1	. . .	2	1	<1	. . .
		IV	1§	. . .	3	1	<1	. . .
		V	1	. . .	2	1	<1	. . .

*V = patients with visceral involvement (other than skin and vascular).
†Includes one patient who had five systems involved but had experienced no Raynaud's phenomenon.
‡Includes three patients with CREST syndrome, duration three, eight and 20 years, organ involvlement three systems each; three patients without Raynaud's phenomenon.
§Patient with Raynand's phenomenon.

respect to capillary class. No patient in the study group had inflammatory joint disease, and articular system involvement was not evaluated.

RESULTS

Study Group. The main finding in the study group was the presence of clusters of dilated capillary loops as described earlier [19,20]. These capillary findings were divided into the following five classes (Figures 1 and 2). Class I: Normal or slightly dilated capillary loops only. Class II: Definitely dilated loops confined to nailfold-distal phalanx. Class III: Extremely dilated loops confined to nailfold-distal phalanx. Class IV: Definitely dilated loops on nailfold-distal phalanx and definitely dilated loops found on at least one site other than nailfold-distal phalanx. Class V: Extremely dilated loops on nailfold-distal phalanx and extremely dilated or definitely dilated loops found at one site other than nailfold-distal phalanx.

The clinical features and the capillary class distri-

TABLE III System Involvement in Study Group

Class	No. of Patients	Involvement						
		Peripheral Vascular	Cutaneous	Gastro-intestinal	Pulmo-nary	Cardiac	Muscular	Renal
I	8	8	2	1	1	1	. . .	. . .
II*	7	7	2	2	1	. . .	. . .	. . .
III†	4	3	3	2	2	1	2	. . .
IV‡	9	9	9	6	4	4	1	. . .
V§	16	13	14	11	11	7	2	2
Total	44	40	30	22	19	13	5	2
Per cent	100	91	68	50	43	30	11	7

*Three had only vascular involvement, one had vascular and cutaneous involvement, one had vascular plus gastrointestinal involvement, one had vascular and pulmonary, and a seventh had vascular, cutaneous and gastrointestinal involvement.
†One had dermatomyositis with pulmonary involvement, and the remaining three had scleroderma. Of these three, one had vascular, cutaneous and gastrointestinal involvement; one had vascular, cutaneous, gastrointestinal and muscular involvement; and one had vascular, cutaneous, cardiac and pulmonary involvement.
‡All patients had vascular and cutaneous involvement. In eight, more than these two systems were involved; three had gastrointestinal, pulmonary and cardiac involvement; one had gastrointestinal and pulmonary involvement; two had gastrointestinal involvement; and one each had cardiac or muscle involvement.
§Of these 16 patients, three denied any history of Raynaud's phenomenon. One patient had dermatomyositis with muscular and cardiac involvement. In seven patients three systems were involved: three had cutaneous, vascular and gastrointestinal involvement; two had cutaneous, vascular and pulmonary involvement; one had cutaneous, gastrointestinal and pulmonary involvement; and one had vascular, gastrointestinal and cardiac involvement. Five patients had four systems involved: three had cutaneous, vascular, gastrointestinal and pulmonary involvement; one had cutaneous, vascular, pulmonary and cardiac involvement; and one had cutaneous, gastrointestinal, pulmonary and cardiac involvement. Two patients had five systems involved: cutaneous, vascular, pulmonary, cardiac and either gastrointestinal or renal. One patient had all seven systems involved and died with malignant hypertension a week after examination. A second patient in this class with four systems involved died at another hospital with congestive heart failure, thought to be secondary to a viral infection of the respiratory tract, four months after examination.

bution among diagnostic subgroups of the 44 study patients are shown in Table II.

In the study group (Table III) 36 of 44 patients had abnormal capillary patterns (scleroderma = 26 of 28, Raynaud's syndrome = seven of 13, dermatomyositis = three of three). Furthermore, capillary class was correlated with organ involvement in the study patients (Figure 3 and Table IV). The details of patients with abnormal capillary patterns (classes II through V) are given in footnotes to Table III.

Eight study patients had class I capillary patterns (Table III). Of these eight, six had only vascular involvement, a seventh had vascular and cutaneous involvement, and the eighth had vascular, cutaneous, gastrointestinal, pulmonary and cardiac involvement, and died of malignant hypertension several months after examination, an exception to the study.

It was possible to reexamine 18 of the 44 study patients, eight with Raynaud's syndrome, nine with scleroderma and one with dermatomyositis. Follow-up observations performed nine to 22 months after the initial observations were not used in the data analyzed. Of the eight patients with Raynaud's syndrome, four showed no change in capillary class or in clinical condition. Two showed a decrease in capillary class (toward normal) with no obvious clinical change. One showed a change from class II to IV and, unfortunately, was lost to clinical evaluation. The eighth patient with Raynaud's syndrome had the most abnormal capillary findings (class IV) in the entire group with Raynaud's syndrome *and* these changed from class IV to V. In this patient pericarditis with effusion developed during the period of follow-up observation.

Of the nine patients with scleroderma, capillary class and clinical condition remained unchanged in seven. The eighth patient showed a change from class II to IV with no change in clinical condition over 21 months. The ninth patient showed a change from class V to class IV with no clinical change.

The single patient with dermatomyositis in whom repeated observations were made showed a change from class IV during active myositis to class II during a remission associated with therapy to class V during an exacerbation of her disease. Of the five patients of the study group who died during the study, class V capillary changes were observed in four (one dermatomyositis, three scleroderma) and class I capillary changes (scleroderma) in one, a previously mentioned exception to the study conclusion.

Control Groups. Group 1: Capillary findings in the rheumatic control group were in agreement with the pattern abnormalities reported previously [19,20]. Of the nine patients with rheumatoid arthritis, three showed prominent subpapillary plexus with normal vessel size in the nailfold. Four more of these revealed a plexus in the area of the dorsal skin proximal to the fourth metacarpal-phalangeal joint, a variation also seen in normal subjects. Two had no significant findings. One of the two patients with psoriatic arthritis had prominent subpapillary plexus, the second was normal. Three of seven patients with systemic lupus erythematosus showed characteristic punched-out, window lesions with loss of capillaries and prominence of subpapillary vessels in affected skin. The others showed no significant abnormalities. In one patient with Sjögren's syndrome only long, somewhat dilated loops were revealed but no visible plexus. In only one patient of this control group, a patient with periarteritis nodosa, were several unusual, large clusters of widely dilated capillary loops seen in numerous grossly visible telangiectatic spots on her hands, face and legs.

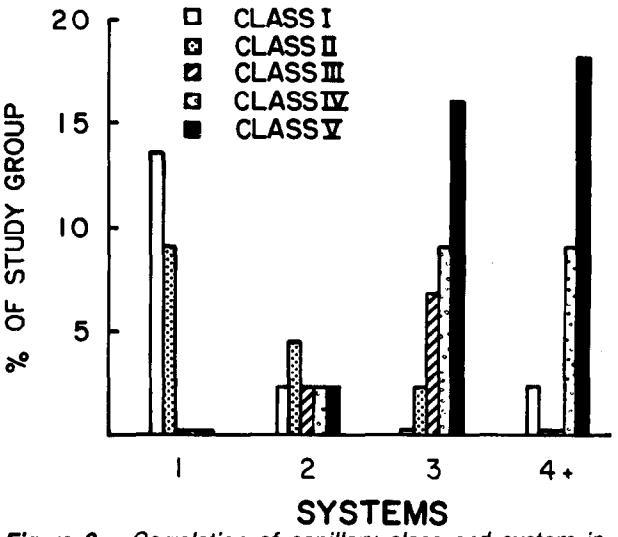

Figure 3. *Correlation of capillary class and system involvement in scleroderma, Raynaud's syndrome and dermatomyositis.*

Group 2: In the hospitalized control group no markedly dilated loops were seen. Ten patients in this group had some visible subpapillary plexus in the nailfold, a nonspecific finding.

Group 3: Of the 20 healthy volunteers, one woman aged 59 had several definitely dilated loops and some loss of capillaries surrounding these in nailfold-distal phalanx

TABLE IV Correlation Between Capillary Class and System Involvement in the Study Group

No. of Systems Involved	Capillary Class		X^2*	P
	I–II	III–V		
1 or 2	13	3	21.72	<0.001
3 or more	2	26		

*With Yates correction.

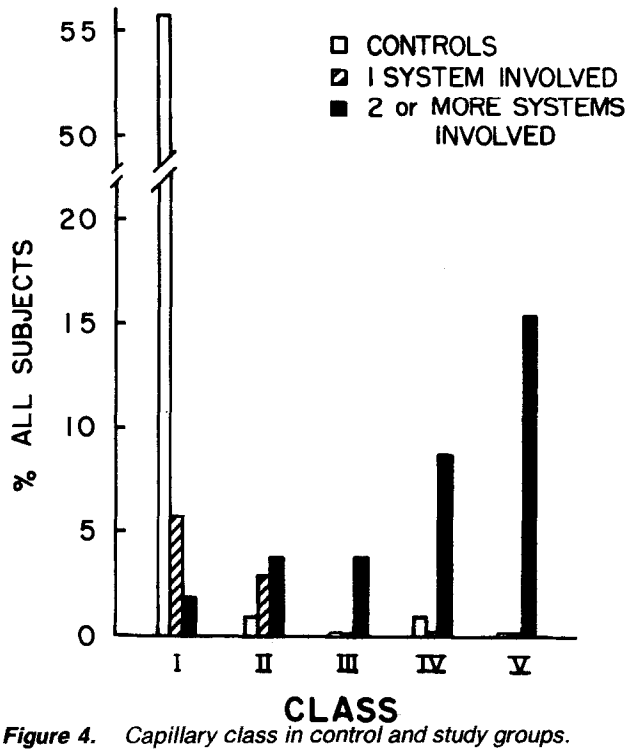

Figure 4. Capillary class in control and study groups.

(class II). This patient was asymptomatic and no further evaluation was made. All other control subjects were normal, with three patients revealing subpapillary plexus in the nailfold.

Group Comparisons. Figure 3 illustrates the significant correlation between capillary class (defined herein) and number of systems involved ($p < 0.001$, Table IV). The comparison of capillary findings between study group and control groups, shown in Figure 4 and Table V, disclosed a significant difference ($p < 0.001$) in the prevalence of abnormal capillary patterns in these groups of subjects.

Since Raynaud's phenomenon and gastrointestinal involvement are the most frequently observed features in addition to the skin lesions in scleroderma, and since these three systems are so frequently involved together in these patients [25], the relationship between capillary abnormalities and system involvement was examined after excluding these three most commonly affected systems. Table VI demonstrates that the degree of capillary abnormality is closely associated with visceral involvement after skin, vascular and gastrointestinal involvement are excluded ($p < 0.001$).

No relationship was found between capillary class and duration of illness (calculated from the onset of Raynaud's phenomenon and/or from the onset of skin involvement).

To remove possible observational bias, the relationship between capillary class and system involvement was also analyzed using data obtained from coded photographs. The results remained similar to the directly observed data, with p values <0.001. The correlation was high between capillary class assignments of subjects based on direct observations and those based on coded photographic data of the same subjects rated "blindly" by a different investigator (Spearman's rank correlation 0.91; $p < 0.0005$).

TABLE V Comparison of Study Group and Control Subjects

Subjects	Capillary Class I	Capillary Class II–V	X^2*	P
Study group	8	36	64.07	<0.001
Control groups	58	2		

*With Yates correction.

TABLE VI Correlation Between Capillary Class and the Type of System Involvement

Capillary Class	Vascular, Cutaneous and/or Gastrointestinal Involvement Only (no. of patients)	Additional System Involvement* (no. of patients) 1	2	3	4	X^2†	P
I	7	...	1	...	...		
II	6	1	...	...	...		
III	1	1	2	...	...	13.16	<0.001
IV	3	3	3	...	...		
V	3	7	4	1	1		

*In a total of 24 patients.

†X^2 between capillary classes I and II versus III, IV, V and the three common systems versus additional system involvement; with Yates correction.

COMMENTS

In the present study, patients with scleroderma, Raynaud's syndrome and dermatomyositis were considered as a single study group for the following reasons: (1) distinctive and similar skin capillary patterns had been previously observed in these disorders [19–21]; (2) the small artery lesion found in these three disorders is often indistinguishable; and (3) clinically there is an overlap, generally in the early stages, among these three disorders [1].

Concerning the second point, the small artery lesion in scleroderma has been well characterized in several viscera, especially the kidney [7], the lung [26] and, more recently, the heart [26,27]. Many classic studies of Raynaud's disease (i.e., Raynaud's phenomenon without connective tissue disease or other systemic diagnosis) have shown small artery changes similar to those seen in scleroderma [2]. More recently, patients with dermatomyositis have been shown to have small artery lesions similar to those in scleroderma [28]. Therefore, in the present investigation designed to study the association between the small artery (on the basis of organ involvement) and capillary lesions, these three overlapping disorders with similar small artery abnormalities were combined.

The results of this study confirm and extend our previous findings of characteristic microvascular abnormalities present in the finger skin of patients with scleroderma and dermatomyositis [19,20].

In addition, the results suggest that the abnormal microvascular patterns are not exclusively associated with local skin disease but reflect systemic involvement and can be seen in some patients in the absence of clinically observable skin disease. The data also suggest that capillary microscopy may be helpful for early recognition of scleroderma among subjects with Raynaud's syndrome. First, in the Raynaud's syndrome group, all subjects with involvement other than skin and vascular system had abnormal capillaries of class II or higher. Furthermore, in the patient with the highest capillary class, pericarditis and other features suggestive of scleroderma developed at follow-up, and her capillary class increased from IV to V. Secondly, in contrast, all six patients with Raynaud's phenomenon alone had class I capillaries. Follow-up examinations which were carried out in two of the six showed no change in capillary class or clinical condition. These individual observations warrant a systematic study of a larger group of patients to determine the diagnostic and prognostic role of widefield capillary observations.

Earlier workers have already suggested examination of nailfold capillaries as a useful tool [29,30], but technical problems with quantitation and photography have discouraged the use of clinical capillaroscopy. It has also been difficult to make serial observations of exactly the same area.

Current clinical practice in detecting organ involvement in Raynaud's syndrome and scleroderma consists of either the study of symptoms with standard physical and laboratory examinations or the elaborate, invasive and occasionally harmful exploration of subtle organ involvement by investigative technics. Repeated use of these investigative technics is not feasible for ethical, economic and personal reasons, unless of course new clinical features justify such procedures. Widefield capillary microscopy, on the other hand, is an inexpensive and noninvasive test which can be repeated at any desired interval. The present use of widefield microscopy and photomicrography permits ready site relocation and capillary pattern detection.

The close association between the degree of nailfold capillary abnormality and organ involvement in scleroderma is strong support for a generalized vascular pathogenesis for this disorder, as well as confirmation that the capillary abnormalities are a direct mirror of this generalized vascular derangement. Independent lines of investigation have delineated a capacity of scleroderma tissue cells to synthesize connective tissue elements, especially collagen, in increased amounts [31]. The connection between the microvascular and the biosynthetic abnormalities is uncertain. Two hypotheses could be considered: one would assume that a local primary lesion occurs at the endothelial level (which may itself be secondary to a blood-borne agent) which, in turn (together with other factors), leads to an activated state of fibroblast function. The second would ascribe a primary role to the activated fibroblast state and would attempt to understand the microvascular alterations as secondary to a generalized mesenchymal cell defect.

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