European multicentre study to define disease activity criteria for systemic sclerosis.*
I. Clinical and epidemiological features of 290 patients from 19 centres


*Members of the European Scleroderma Study Group are given in the Appendix.

Abstract

Objective—To investigate the existence of differences among European referral centres for systemic sclerosis (SSc) in the pattern of attendance and referral and in the clinical and therapeutical approaches. Methods—In 1995 the European Scleroderma Study Group initiated a multicentre prospective one year study whose aim was to define the disease activity criteria in SSc. During the study period each participating European centre was asked to enrol consecutive patients satisfying American College of Rheumatology criteria for SSc and to fill out for each of them a standardised clinical chart. Patients from various centres were compared and differences in epidemiological, clinical, and therapeutical aspects were analysed. Results—Nineteen different medical research centres consecutively recruited 290 patients. The patients could be divided into two subgroups: 173 with the limited (ISSc) and 117 with the diffuse (dSSc) form of the disease. The clinical and serological findings for the series of 290 patients seemed to be similar to data previously reported. However, when the data were analysed to elicit any differences between the participating centres, a high degree of variability emerged, in both epidemiological and clinical features and in the diagnostic and therapeutic approaches to the disease. Conclusions—The clinical approach to SSc, not only in different countries but also in different centres within the same country, is not yet standardised. To overcome this problem, it will be necessary for the scientific community to draw up a standardised procedure for the management of patients with SSc. This would provide a common research tool for different centres engaged in research on this complex disease.

(Systemic sclerosis (SSc) is a multisystem connective tissue disorder characterised by widespread microvascular and macrovascular damage, and by fibrosis of the skin and internal organs, particularly the gut, lung, heart, and kidney. The extent of skin and internal organ involvement, and the severity and course of the disease may vary greatly. Moreover, the treatment for patients with SSc has not yet been standardised. Thus the clinical approach may differ widely not only between doctors at outpatient clinics and those working in research oriented tertiary centres but also between centres of the same type.

In 1995 the European Scleroderma Study Group was formed and began a multicentre study whose aim was to define a valid set of criteria for disease activity in SSc. Data were gathered on a large number of patients consecutively recruited at 19 different medical research centres in Europe. In this paper we report and discuss the preliminary data gathered at the first observation on 290 patients. In...
Interstitial lung disease: From the onset of the first non-Raynaud's manifestation. Systemic sclerosis (SSc)

Table 3 Definition of some of the items recorded in the clinical chart

<table>
<thead>
<tr>
<th>Item</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Skin score</td>
<td>recorded according to Kahaleh et al. This score is evaluated on a four point scale (0 for normal skin, 1 for thickened skin, 2 for thickened, unpinchable skin, 3 for hidebound skin) at 22 body regions (maximal value 66)</td>
</tr>
<tr>
<td>Oesophageal hypomotility</td>
<td>radiologically documented hypomotility</td>
</tr>
<tr>
<td>Intestinal lung disease</td>
<td>signs of lung fibrosis documented by chest x-ray and/or computed tomography (CT) scan. The standard chest x-ray was scored 0–3: 0 = absence of interstitial lung disease; 1 = bibasilar lung fibrosis; 2 = diffuse interstitial lung fibrosis; 3 = honeycombing. CT was scored 0–3: 0 = absence of interstitial lung disease; 1 = ground glass appearance; 2 = subpleural septal lines; 3 = honeycombing. Test of pulmonary function (forced vital capacity) and carbon monoxide transfer factor were performed, as well</td>
</tr>
</tbody>
</table>

Table 4 Main demographic features of the series of 290 patients with systemic sclerosis

<table>
<thead>
<tr>
<th>Disease Subset</th>
<th>Limited (dSSc)</th>
<th>Diffuse (lSSc)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of Patients</td>
<td>173</td>
<td>117</td>
</tr>
<tr>
<td>F/M</td>
<td>6.2</td>
<td>4.3</td>
</tr>
<tr>
<td>Age at enrolment (years)</td>
<td>55 (13) (range 8–87)</td>
<td>44 (14) (range 5–80)</td>
</tr>
<tr>
<td>Mean disease duration at enrolment (years)*</td>
<td>11 (10) (range 0–56)</td>
<td>8 (7) (range 0–51)</td>
</tr>
<tr>
<td>Time from disease onset to admission (years)‡</td>
<td>8 (9) (range 0–56)</td>
<td>3 (4) (range 0–23)</td>
</tr>
</tbody>
</table>

*From the onset of Raynaud’s phenomenon. †From the onset of the first non-Raynaud’s manifestation. ‡That is, the first visit to the centre.

Table 5 Main demographic features for the two disease subsets, limited and diffuse systemic sclerosis (SSc)

<table>
<thead>
<tr>
<th>Disease Subset</th>
<th>Limited (dSSc)</th>
<th>Diffuse (lSSc)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of Patients</td>
<td>173</td>
<td>117</td>
</tr>
<tr>
<td>F/M</td>
<td>6.2</td>
<td>4.3</td>
</tr>
<tr>
<td>Age at enrolment (years)</td>
<td>55 (range 21–87)</td>
<td>44 (range 8–86)</td>
</tr>
<tr>
<td>Mean disease duration at enrolment (years)*</td>
<td>13 (range 0–56)</td>
<td>8 (range 0–47)</td>
</tr>
</tbody>
</table>

*From the onset of Raynaud’s phenomenon. †From the first non-Raynaud’s manifestation.

Materials and methods

Nineteen centres located in 11 different European countries participated in this study. Each investigator was asked to enrol a consecutive number of patients with SSc, satisfying the preliminary American College of Rheumatology (ACR, formerly American Rheumatism Association) criteria for the classification of the disease.

CLINICAL CHART

A standardised clinical chart was prepared and one copy was sent to each participating centre. Table 1 summarises the demographic and epidemiological information, and table 2 the clinical, laboratory, and other parameters (radiological, etc) that were to be gathered and recorded for each patient.

The chart was made up of four sections. In section I, for each patient the participants were asked to provide demographic information and a patient history; an evaluation based on the preliminary criteria for SSc of the ACR; a classification of the patient’s disease as diffuse or limited (dSSc or lSSc) according to Le Roy et al.

At entry, each patient also had to undergo a complete evaluation, and the results (including symptoms, signs, laboratory and other diagnostic test findings) were to be recorded in section II. This section was divided into 11 parts, focusing on specific categories of disease manifestations (generalised complaints; cutaneous manifestations; vascular manifestations; cardiopulmonary manifestations; articular/muscular manifestations; oculart manifestations; gastrointestinal manifestations; haematological alterations; renal alterations; and neuropsychiatric manifestations), and the 11th on aspecific inflammation and immunological features. At the end of each of the 11 subsections, an additional item was included (A-factor) designed to note any worsening in the relevant manifestation during the month preceding enrolment (according to the patient’s report). Sections III and IV were analogous to section II (except that the A-factor recorded any change in the manifestation compared with the previous observation) and were to be completed for each patient at two later time points—that is, after six and 12 months, respectively. Finally, at the end of sections II–IV, the treatment prescribed...
European multicentre study to define disease activity criteria for systemic sclerosis. I.

Figure 2. Cumulative prevalence of organ involvement based on data from the patient history section—that is, collected at the time of entry into the study.

Table 6. Main clinical and serological features for the case series (at the time of the first observation).

<table>
<thead>
<tr>
<th>Feature</th>
<th>No (%)</th>
<th>Number missing</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Generalised complaints</td>
<td>163/290 (56)</td>
<td>0</td>
</tr>
<tr>
<td>2 Joints/tendons</td>
<td>39/290 (13)</td>
<td>0</td>
</tr>
<tr>
<td>Tendon rubs</td>
<td>17/290 (6)</td>
<td>0</td>
</tr>
<tr>
<td>Myositis</td>
<td>62/290 (21)</td>
<td>0</td>
</tr>
<tr>
<td>Myalgia</td>
<td>69/290 (24)</td>
<td>0</td>
</tr>
<tr>
<td>Muscle weakness</td>
<td>151/290 (52)</td>
<td>0</td>
</tr>
<tr>
<td>Arthralgia</td>
<td>51/290 (18)</td>
<td>0</td>
</tr>
<tr>
<td>Arthritis</td>
<td>79/290 (27)</td>
<td>0</td>
</tr>
<tr>
<td>Flexion contractures</td>
<td>61/290 (21)</td>
<td>0</td>
</tr>
<tr>
<td>Heart</td>
<td>157/290 (54)</td>
<td>0</td>
</tr>
<tr>
<td>Skin</td>
<td>274/290 (94)</td>
<td>0</td>
</tr>
<tr>
<td>Sclerosis</td>
<td>100/290 (34)</td>
<td>0</td>
</tr>
<tr>
<td>Telangiectasia</td>
<td>193/290 (67)</td>
<td>0</td>
</tr>
<tr>
<td>Calcinosis</td>
<td>76/290 (26)</td>
<td>0</td>
</tr>
<tr>
<td>Atrophic skin ulcers</td>
<td>117/290 (40)</td>
<td>0</td>
</tr>
<tr>
<td>Vascular lesions</td>
<td>19/290 (7)</td>
<td>0</td>
</tr>
<tr>
<td>Sicca syndrome*</td>
<td>77/290 (27)</td>
<td>0</td>
</tr>
<tr>
<td>Sicca syndrome†</td>
<td>165/290 (57)</td>
<td>0</td>
</tr>
<tr>
<td>Lung</td>
<td>179/290 (62)</td>
<td>0</td>
</tr>
<tr>
<td>Dyspnoea</td>
<td>127/171 (74)</td>
<td>119</td>
</tr>
<tr>
<td>Intestinal involvement</td>
<td>84/132 (64)</td>
<td>158</td>
</tr>
<tr>
<td>CT scan</td>
<td>89/105 (85)</td>
<td>185</td>
</tr>
<tr>
<td>Reduced TLCO‡</td>
<td>144/285 (51)</td>
<td>5</td>
</tr>
<tr>
<td>Reduced FVC¶</td>
<td>69/252 (27)</td>
<td>38</td>
</tr>
<tr>
<td>Heart</td>
<td>61/290 (21)</td>
<td>0</td>
</tr>
<tr>
<td>Heart failure</td>
<td>9/290 (3)</td>
<td>0</td>
</tr>
<tr>
<td>Peripheral vascular system</td>
<td>53/290 (18)</td>
<td>0</td>
</tr>
<tr>
<td>Hypertension</td>
<td>266/290 (92)</td>
<td>0</td>
</tr>
<tr>
<td>Digital pitting</td>
<td>181/290 (62)</td>
<td>0</td>
</tr>
<tr>
<td>Digital necrosis</td>
<td>50/290 (17)</td>
<td>0</td>
</tr>
<tr>
<td>Gastrointestinal apparatus</td>
<td>229/290 (79)</td>
<td>0</td>
</tr>
<tr>
<td>Oesophageal involvement</td>
<td>31/290 (11)</td>
<td>0</td>
</tr>
<tr>
<td>Diarrhoea</td>
<td>52/290 (18)</td>
<td>0</td>
</tr>
<tr>
<td>Kidney involvement¶</td>
<td>23/290 (8)</td>
<td>0</td>
</tr>
<tr>
<td>Nervous system</td>
<td>25/290 (9)</td>
<td>0</td>
</tr>
<tr>
<td>Headache/trigeminal neuropathy</td>
<td>44/290 (15)</td>
<td>0</td>
</tr>
<tr>
<td>Depression</td>
<td>233/249 (94)</td>
<td>41</td>
</tr>
<tr>
<td>ANA*</td>
<td>57/253 (23)</td>
<td>37</td>
</tr>
<tr>
<td>ACA*</td>
<td>116/249 (43)</td>
<td>21</td>
</tr>
<tr>
<td>Scl-70</td>
<td>22/215 (10)</td>
<td>75</td>
</tr>
<tr>
<td>aCL¶</td>
<td>53/231 (23)</td>
<td>59</td>
</tr>
<tr>
<td>ESR*</td>
<td>71/273 (26)</td>
<td>17</td>
</tr>
<tr>
<td>CRP¶</td>
<td>34/239 (14)</td>
<td>51</td>
</tr>
<tr>
<td>LDH¶</td>
<td>23/210 (11)</td>
<td>80</td>
</tr>
<tr>
<td>CK¶</td>
<td>18/234 (9)</td>
<td>56</td>
</tr>
<tr>
<td>Low C3/C4</td>
<td>33/233 (14)</td>
<td>57</td>
</tr>
</tbody>
</table>

*Both xerostomia and xerophthalmia.
†Either pericardial involvement or arrhythmias or conduction defects.
‡Either proteinuria or haematuria or increased creatininemia.
¶CT = computed tomography; TLCO = carbon monoxide transfer factor; FVC = forced vital capacity; ANA = antinuclear antibodies; ACA = anticientromere antibodies; aCL = anticardiolipin antibodies; RF = rheumatoid factor; ESR = erythrocyte sedimentation rate; CRP = C reactive protein; LDH = lactate dehydrogenase; CK = creatine kinase.

Results

DEMOGRAPHIC AND HISTORICAL DATA

Table 4 summarises the main demographic features of the 290 patients recruited for this study. Women outnumbered men by 5.3 to 1, and there was a wide variation in the epidemiological parameters between patients. The disease duration, defined as the time elapsed from the onset of the first symptom (usually Raynaud’s phenomenon), ranged from 0 to 56 years. When defined as the time elapsed from the first non-Raynaud manifestation, it ranged from 0 to 51 years. Thus a high percentage of the patients had disease of long duration. There was also in most cases a significant lapse in time between the patient’s admission to the participating centre and his/her enrolment in the study; 190/290 had already been followed up at the respective centres for at least one year before being recruited into the study.

As a result of the recruitment process, all the 290 patients fulfilled the ACR criteria for SSC (165 patients met the major criterion and 125 met two of the three minor criteria). One hundred and seventeen had the diffuse and 173 the limited form of the disease. Table 5 shows the demographic features of the patients by disease subset. A significant difference was seen in the mean disease duration, when evaluated from the onset of Raynaud’s phenomenon (13 ± 8 years), and in the mean age at enrolment (55 ± 49 years) between patients with ISSc and dSSc.

An analysis of variance showed a significant variation between centres in epidemiological parameters (fig 1), including the age at enrolment, the disease duration from the onset of the Raynaud’s phenomenon, and the male to female ratio, which ranged from 1/2 to 1/20. In addition, significant differences emerged in...
subset distribution, the ISSc to dSSc ratio ranging from 5/1 to 1/2.5.

Figure 2 shows the prevalence of organ involvement as reported in the patients’ histories collected at the time of entry into the study. Significant differences (data not shown) emerged in the prevalence of various disease manifestations—namely, gut, lung, heart, kidney, joint, and muscle involvement, while the prevalence of skin sclerosis and Raynaud’s phenomenon showed a slight difference only. Of course, a number of these differences must be ascribed to different investigative methods. Nevertheless, the detected differences in sex, age, subset distribution, and clinically detectable manifestations, such as joint involvement, point to actual diversities among different centre series.

CLINICAL AND SEROLOGICAL FEATURES AT THE TIME OF ENROLMENT

Table 6 shows the clinical features reported for the 290 patients at enrolment. The most common findings were skin sclerosis, Raynaud’s phenomenon, interstitial lung involvement, and oesophageal involvement. Skin sclerosis was found in 274 patients. Therefore, about 6% of the cases could be diagnosed as SSc without sclerodera. Kidney involvement (that is, either proteinuria or haematuria or increased creatininaemia) was detected in 52/290 (18%) of the patients, but no cases of renal crisis were recorded. Table 6 clearly shows that several items were characterised by a high number of missing values. This was particularly true for investigations to detect internal organ involvement. Despite these limitations, however, the figures for clinically detectable manifestations, such as scleredema, melanoderma, arthritis etc, must be considered reliable, each observer being an experienced clinician in scleroderma to whom clear cut guidelines had been provided.

When patients were compared by disease subset (dSSc vs ISSc), statistically significant differences in some of the clinical and serological manifestations were found (table 7; data shown in italics). Patients with dSSc had more severe skin involvement, associated with a higher frequency of functional impairment (that is, flexion contractures), tendon friction rubs, melanoderma, acro-osteolysis, digital necrosis, and reduced forced vital capacity and carbon monoxide transfer factor.

An analysis was carried out to elicit any differences between the centres in the prevalence of clinically detectable manifestations. A high degree of variability was found among centres in the reported frequency of various disease manifestations (figs 3 and 4). On the whole, the greatest variation was seen in the parameters of scleredema, melanoderma, calcinosis, and digital infarcts. As already stated, these items had been so carefully defined and actually are so easy to detect that the differences which emerged are to be considered true. On the other hand, the differences which emerged in the prevalences of interstitial lung involvement, oesophageal involvement, and autoantibody profile may depend on differing investigative tools.

**Table 7** Prevalence of the different clinical and serological features among the two disease subsets

<table>
<thead>
<tr>
<th>Manifestation (%)</th>
<th>ISSc</th>
<th>dSSc</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Generalised complaints</td>
<td>53</td>
<td>60</td>
</tr>
<tr>
<td>2 Joints/tendons</td>
<td>70</td>
<td>88</td>
</tr>
<tr>
<td>Tendon rubs</td>
<td>6</td>
<td>23</td>
</tr>
<tr>
<td>Myositis</td>
<td>5</td>
<td>6</td>
</tr>
<tr>
<td>Muscle weakness</td>
<td>20</td>
<td>24</td>
</tr>
<tr>
<td>Arthritis</td>
<td>21</td>
<td>27</td>
</tr>
<tr>
<td>Arthralgia</td>
<td>51</td>
<td>60</td>
</tr>
<tr>
<td>Arthritis</td>
<td>17</td>
<td>17</td>
</tr>
<tr>
<td>Flexion contractures</td>
<td>16</td>
<td>46</td>
</tr>
<tr>
<td>Joint contractures</td>
<td>16</td>
<td>29</td>
</tr>
<tr>
<td>3 Skin</td>
<td>100</td>
<td>100</td>
</tr>
<tr>
<td>Scleredema</td>
<td>56</td>
<td>51</td>
</tr>
<tr>
<td>Skin sclerosis</td>
<td>94</td>
<td>95</td>
</tr>
<tr>
<td>Melanoderma</td>
<td>23</td>
<td>51</td>
</tr>
<tr>
<td>Calcinosis</td>
<td>65</td>
<td>68</td>
</tr>
<tr>
<td>Calcinosis</td>
<td>20</td>
<td>22</td>
</tr>
<tr>
<td>Atrophic skin ulcers</td>
<td>38</td>
<td>43</td>
</tr>
<tr>
<td>Vascular lesions</td>
<td>5</td>
<td>9</td>
</tr>
<tr>
<td>Sicca syndrome*</td>
<td>28</td>
<td>25</td>
</tr>
<tr>
<td>4 Heart/lung</td>
<td>90</td>
<td>93</td>
</tr>
<tr>
<td>Dyspnoea</td>
<td>58</td>
<td>67</td>
</tr>
<tr>
<td>Intestinal involvement (x ray)</td>
<td>67</td>
<td>72</td>
</tr>
<tr>
<td>Reduced FVC†</td>
<td>16</td>
<td>44</td>
</tr>
<tr>
<td>Reduced TLC†</td>
<td>45</td>
<td>58</td>
</tr>
<tr>
<td>Heart failure</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>5 Peripheral vascular system</td>
<td>96</td>
<td>97</td>
</tr>
<tr>
<td>Arterial hypertension</td>
<td>19</td>
<td>17</td>
</tr>
<tr>
<td>Raynaud’s phenomenon</td>
<td>92</td>
<td>91</td>
</tr>
<tr>
<td>Digital pitting</td>
<td>62</td>
<td>62</td>
</tr>
<tr>
<td>Digital necrosis</td>
<td>13</td>
<td>22</td>
</tr>
<tr>
<td>6 Gastrointestinal apparatus</td>
<td>76</td>
<td>81</td>
</tr>
<tr>
<td>Diarrhoea</td>
<td>11</td>
<td>10</td>
</tr>
<tr>
<td>Nervous system</td>
<td>26</td>
<td>19</td>
</tr>
<tr>
<td>Headache/trigeminal neuropathy</td>
<td>11</td>
<td>5</td>
</tr>
<tr>
<td>Depression</td>
<td>16</td>
<td>15</td>
</tr>
<tr>
<td>8 Routine laboratory tests</td>
<td></td>
<td></td>
</tr>
<tr>
<td>ESR &gt;30 mm/1st h</td>
<td>25</td>
<td>27</td>
</tr>
<tr>
<td>High CRP‡</td>
<td>11</td>
<td>19</td>
</tr>
<tr>
<td>High LDH‡</td>
<td>10</td>
<td>12</td>
</tr>
<tr>
<td>High CK‡</td>
<td>7</td>
<td>9</td>
</tr>
<tr>
<td>Low Cr/C××</td>
<td>13</td>
<td>16</td>
</tr>
</tbody>
</table>

*Both xerostomia and xerophthalmia.
†FVC = forced vital capacity; TLC = carbon monoxide transfer factor; ESR = erythrocyte sedimentation rate; CRP = C reactive protein; LDH = lactic dehydrogenase; CK = creatine kinase.

When patients were compared by disease subset (dSSc vs ISSc), statistically significant differences in some of the clinical and serological manifestations were found (table 7; data shown in italics). Patients with dSSc had more severe skin involvement, associated with a higher frequency of functional impairment (that is, flexion contractures), tendon friction rubs, melanoderma, acro-osteolysis, digital necrosis, and reduced forced vital capacity and carbon monoxide transfer factor.

An analysis was carried out to elicit any differences between the centres in the prevalence of clinically detectable manifestations. A high degree of variability was found among centres in the reported frequency of various disease manifestations (figs 3 and 4). On the whole, the greatest variation was seen in the parameters of scleredema, melanoderma, calcinosis, and digital infarcts. As already stated, these items had been so carefully defined and actually are so easy to detect that the differences which emerged are to be considered true. On the other hand, the differences which emerged in the prevalences of interstitial lung involvement, oesophageal involvement, and autoantibody profile may depend on differing investigative tools.

**TREATMENT**

Figure 5 shows the treatment prescribed by the participating clinicians at the time of the first...
The aim of this multicentre European study, which is still continuing, will be to develop a valid set of disease criteria for SSc based on clinical data collected in a standardised manner on a large number of patients. Here we report the results of our analysis of the data gathered during the first phase of this study (patient history and evaluation at enrolment). The epidemiological and clinical serological findings for our series of 290 patients seem to be similar to data previously reported by others (tables 8 and 9). However, the data furnished by the 19 centres were far from homogeneous. The complex picture presented by SSc, characterised by a high variability in its clinical manifestations and treatment modalities, has already been discussed by other authors, though not in the light of rigorously designed, large scale studies.

Our prospective study, in which all of the centres were required to follow the same clearly defined protocol, clearly demonstrates the magnitude of the problem presented by this disease. The epidemiological variability observed as well as the high variability in the reported incidence of clinical features (such as calcinosis, sclerodema, scleroderma, telangiectasia) may be ascribed to differences in the nature of the participating centres. It would be hard to believe that experienced clinicians might have failed to detect single manifestations such as melanoderma, digital necrosis, etc. On the other hand, the high number of missing values concerning most investigative methods prevents us from discussing the differences detected in the internal organ involvement. Nevertheless, the absence of patients with scleroderma renal crisis in our series deserves some comments. This complication has been identified as one of the leading causes of death in the North American series. However, studies conducted in Europe seem to indicate a lower prevalence. The absence of such a manifestation in our series may depend, however, on the nature of the study protocol, which enrolled consecutive patients and therefore might have excluded more severe cases. As far as kidney disease is concerned, we detected either proteinuria or haematuria or increased creatininemia in 18% of our patients with SSc. However, we cannot confidently ascribe these alterations to SSc itself, because we did not correct the data for the presence of diabetes, hypertension, and other causes of these alterations.

In conclusion, certain observations may be made about SSc based on the preliminary results of our multicentre study. Despite our study design—which used a standardised chart that included all of the most generally accepted diagnostic parameters for SSc, carefully defined according to authoritative sources—we still found considerable differences between centres in the use of diagnostic tests and the therapeutic approach. This reflects the as yet unstandardised approach to SSc in different countries. To overcome these problems the scientific community will need to draw up a standardised procedure for the management of patients with SSc. This should include a


**Figure 6** Treatments used by the different centres. In the box plots the 10th, 25th, 50th (median), 75th, and 90th centiles of each variable are shown. Values above the 90th and below the 10th centiles are plotted as points.

Table 8 Comparison of demographic, clinical, and serological data for the present series with similar data from other European series

<table>
<thead>
<tr>
<th>Variable</th>
<th>Present series</th>
<th>Vysyáral et al.</th>
<th>Jacobsen et al.</th>
<th>Nagy and Gázsyári</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>290</td>
<td>184</td>
<td>230</td>
<td>171</td>
</tr>
<tr>
<td>Women (%)</td>
<td>84</td>
<td>86</td>
<td>82</td>
<td>87</td>
</tr>
<tr>
<td>Limited/diffuse SSc (%)</td>
<td>1.5</td>
<td>1.0</td>
<td>2.8</td>
<td>3.75</td>
</tr>
<tr>
<td>Mean age at onset (years)</td>
<td>53</td>
<td>31</td>
<td>44</td>
<td>44</td>
</tr>
<tr>
<td>Raynaud’s phenomenon (%)</td>
<td>92</td>
<td>100</td>
<td>96</td>
<td>95.3</td>
</tr>
<tr>
<td>Digital pitting (%)</td>
<td>62</td>
<td>71</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>Telangiectasia (%)</td>
<td>67</td>
<td>75</td>
<td>73</td>
<td>64.3</td>
</tr>
<tr>
<td>Calcinosis (%)</td>
<td>26</td>
<td>60</td>
<td>42</td>
<td>42.9</td>
</tr>
<tr>
<td>Joint involvement (%)</td>
<td>66</td>
<td>53</td>
<td>18</td>
<td>—</td>
</tr>
<tr>
<td>Scleroderma renal crisis (%)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1.2</td>
</tr>
</tbody>
</table>

Table 9 Comparison of demographic, clinical, and serological data for the present series with similar data from other, non-European countries

<table>
<thead>
<tr>
<th>Variable</th>
<th>Present series</th>
<th>Steen et al.</th>
<th>Chandran et al.</th>
<th>Kayanana et al.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>290</td>
<td>397</td>
<td>115*</td>
<td>275*</td>
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<tr>
<td>Women (%)</td>
<td>84</td>
<td>83</td>
<td>84</td>
<td>86</td>
</tr>
<tr>
<td>Limited/diffuse overlap SSc</td>
<td>1.5/1.0</td>
<td>0.9/1.0</td>
<td>6/1.6</td>
<td>7 : 5.7 : 3</td>
</tr>
<tr>
<td>Mean age at onset (years)</td>
<td>53</td>
<td>42</td>
<td>44</td>
<td>43</td>
</tr>
<tr>
<td>Raynaud’s phenomenon (%)</td>
<td>92</td>
<td>96</td>
<td>96</td>
<td>98</td>
</tr>
<tr>
<td>Digital pitting (%)</td>
<td>62</td>
<td>66</td>
<td>66</td>
<td>25</td>
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<tr>
<td>Telangiectasia (%)</td>
<td>67</td>
<td>82</td>
<td>82</td>
<td>25</td>
</tr>
<tr>
<td>Calcinosis (%)</td>
<td>26</td>
<td>39</td>
<td>39</td>
<td>—</td>
</tr>
<tr>
<td>Joint involvement (%)</td>
<td>66</td>
<td>64</td>
<td>64</td>
<td>35</td>
</tr>
<tr>
<td>Scleroderma renal crisis (%)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>

*SSc was classified in three groups: dc-SSc, lc-SSc, and SSC in overlap.
†SSc was classified in three groups related to the autoantibody profile: dc-SSc, lc-SSc, and SSC in overlap.
European multicentre study to define disease activity criteria for systemic sclerosis. I. Clinical and epidemiological features of 290 patients from 19 centres


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