Supplementary file

Patients' characteristics

Demographic characteristics included age in years, sex and total follow-up duration in the CCISS cohort. Disease duration was defined as months between onset of RP and a first non-RP symptom. Non-RP symptoms were defined as follows: cardiac involvement was based on the clinical diagnosis by a team of expert rheumatologists, pulmonologists and cardiologists. A decreased left ventricular ejection fraction was defined as ≤54% (32) and was estimated using echocardiography by an experienced cardiologist. Pulmonary arterial hypertension (PAH) was defined as a mean pulmonary arterial pressure ≥25 mmHg at rest as assessed by right heart catheterization; including presence of pre-capillary PH, defined by a pulmonary capillary wedge pressure ≤15 mmHg and a pulmonary vascular resistance >3 Wood units on right heart catheterization. Diagnosis of interstitial lung disease (ILD) was determined based on presence of interstitial fibrosis or ground glass opacities on high resolution computerized tomography (HRCT) of the thorax as reported by a radiologist. Moreover, a forced vital capacity or diffusing capacity for carbon monoxide <80% on the pulmonary function test was used to determine if radiologic ILD was clinically meaningful. The severity of skin involvement was measured by means of the modified Rodnan Skin Score (mRSS) (33) and patients were categorized as non-cutaneous, limited cutaneous or diffuse cutaneous. Other non-RP symptoms included puffy fingers or sclerodactyly, fingertip lesions (digital tip ulcers or pitting scars), telangiectasia and abnormal nailfold capillaries.