Supplementary Online Content


eMethods. Further Methodology

eFigure. ROC Curves Assessing the Ability to Discriminate the Presence of PH

This supplementary material has been provided by the authors to give readers additional information about their work.
**Data collection included:**

- **age, sex, and smoking history (yes/no),**
- **disease characteristics: disease duration since the date of the First Non Raynaud’s Phenomenon symptom (FNRP); disease duration since inclusion; scleroderma classification according to Leroy’s classification (limited and diffuse cutaneous form);**¹ autoantibodies status (anti-centromere or ACA, anti-topoisomerase I or ATA and anti-RNA polymerase III or RNAIII antibodies); EUropean Scleroderma Trials and Research (EUSTAR) Activity score;² Medsger severity score;³ and Health Assessment Questionnaire-Disability Index (HAQ-DI);⁴
- **treatments (yes/no): corticosteroid; immunosuppressive (cyclophosphamide, mycophenolate mofetil, mycophenolate acid, azathioprine, methotrexate, anticalcineurin, D-penicillamin, antimalarial synthesis, polyvalent immunoglobulin, anti-CD20, anti-TNFα, or anti-IL6) and PH (phosphodiesterase type 5 inhibitor, endothelin receptor antagonist, prostacyclin analogues, or guanylate cyclase inhibitor) treatments,**
- **cardio-pulmonary parameters: New York Heart Association (NYHA) functional score; 6-minute walk test (6MWT) performed as recommended by the American Thoracic Society (ATS);⁵ interstitial lung disease (ILD) diagnosed on a chest high-resolution CT-scan and staged according to Goh’s criteria⁶; pulmonary function tests (PFT) with total forced vital capacity (FVC), diffusing capacity of the lung for carbon monoxide (DLCO), diffusing coefficient for carbon monoxide (KCO); N-terminal pro-brain natriuretic peptide (Nt-proBNP); precapillary pulmonary hypertension (PH) was defined according to the last guidelines by right heart catheterism (RHC) as a
pulmonary artery pressure (PAPm) ≥25 mmHg, pulmonary artery wedge pressure (PAWP) ≤15 mmHg;\textsuperscript{7} transthoracic echocardiography (TTE) with left ventricular ejection fraction (LVEF), peak tricuspid regurgitation velocity (TRV), estimated systolic pulmonary artery pressure (sPAP) and right atrial area,

- skin parameters: modified Rodnan skin score (mRSS);\textsuperscript{8} digital ulcers (DU) (history and/or presence during the clinical evaluation); Nailfold videocapillaroscopy (NVC) with determination of patterns of microangiopathy using Cutolo stages (early, active and late);\textsuperscript{9} calcinosis was defined by at least one calcification in soft tissues on the clinical examination and/or on X-Rays,

- renal parameters: history of scleroderma renal crisis (SRC); glomerular filtration rate (GFR) estimated by the Modification of Diet in Renal Disease (MDRD) equation; urinary abnormalities (proteinuria and/or haematuria),

- vascular parameters: history of deep vein thrombosis, pulmonary embolism or arterial thrombosis (myocardial infarction, ischemic stroke, or acute limb ischemia),

- musculoskeletal parameters: joint symptoms (pain or swelling); muscle symptoms (pain or weakness),

- biological parameters: anemia defined by haemoglobin ≤13g/dL for male and ≤12g/dL for female gender; ferritin level; C-reactive protein (CRP) and uricemia. Patients were tested for LA, aCL (IgG isotype) and anti-\(\beta\)2GpI (IgG isotype). Lupus anticoagulant was detected in plasma by a dilute Russell’s viper venom time (Siemens), and partial thromboplastin time test (HemosIL Silica Clotting Time Werfen) as screening and confirmation tests with calculating a normalized ratio. Anti-cardiolipine antibodies (aCL) and anti-\(\beta\)2 glycoprotein 1 antiantibodies (anti-\(\beta\)2GpI) were measured using commercial ELISA assays (Orgentec, Trappes, France); positive titer was defined as ≥10 UGPL/mL (aCL) or ≥10 UA/mL (anti-\(\beta\)2GpI).
eReferences.


eFigure 1. ROC curves assessing the ability to discriminate the presence of PH for: (A) the total TA number on the whole body, (B) TA score, (C) the TA number on hands and face, (D) and the TA number on hands.