pulmonary arterial pressure, mPAP ≥25 mmHg) in the absence of significant interstitial lung disease (ILD), based on high resolution computed tomography and lung function tests. Borderline PAH was defined as mPAP of 20–24 mmHg in the absence of significant ILD. Patients with pulmonary hypertension (PH) due to other causes were excluded. Descriptive statistics and logistic regression analyses were performed and tested by the goodness-of-fit test with area under the curve (AUC).

Results: Mean age at onset was 54±14.1, at RHC 61±10.9 years. The time from sera to RHC was 0.9±3.1 years. In total, 73.7% (123) patients had limited cutaneous SSc. 46.7%48 were positive for anti-centromere antibody and 80.2% (134) of females. Of these, 123 (73.7%) patients were included in the study, 28 patients with PAH, 45 borderline PAH and 50 patients with no PH. Mean VEGF-A, C.D, CCL21 and Ang2 levels are shown in figure 1A. VEGF-A was significantly higher in SSc patients compared to healthy controls (p=0.001), no significant differences between PAH and no PH patients were found. VEGF-D was decreased in SSc-PAH but not significantly. CCL21 and Ang2 levels were increased in patients with PAH, whereas in VEGF-C was significantly decreased in patients with PAH (figure 1 B-D). In univariable logistic regression analyses, VEGF-C (OR 0.99, 95% CI 0.997–0.998, p=0.001, AUC=0.79), CCL21 (OR 1, 95% CI 1–1.003, p=0.050, AUC=0.69) and Ang2 (OR 1, 95% CI 1–1.001,p=0.49, AUC=0.67) were associated with PAH compared to no PH patients. Due to few event numbers, no multivariable analyses were performed.

Conclusions: The present study is the first to demonstrate dysregulation of lymphangiogenic factor expression of multiple targets in sera of SSc-PAH patients.

Disclosure of Interest: None declared


FR10432

SINGLE-PORT THORACOSCOPIC SYMPATHICOTOMY FOR TREATMENT RESISTANT RAYNAUD’S PHENOMENON. FIRST REPORT OF A NOVEL MINIMALLY-INVASIVE ENDOSCOPIC TECHNIQUE

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Background: Raynaud’s phenomenon of the hands is a great burden in daily life and reduces quality of life in patients with or without an underlying connective tissue disease. Although vasodilatory treatment may be effective in some patients, complaints may be resistant to treatment, for which additional treatment options are very limited. In earlier years, thoracic sympathectomy by anterior or axillary thoracotomy has been shown effective, but with a great surgical burden and limited durability. In the 90s, endoscopic procedures were introduced. However, these techniques needed multiple endoscopic ports and still performed a relatively mutilating sympathectomy of the sympathetic ganglia. In our centre, the single-port thoracoscopic sympathicotomy (SPTS) was developed, which is a new minimally invasive endoscopic technique with a limited surgical burden.

Objectives: The aim of the current pilot study was to evaluate feasibility and efficacy of SPTS in patients with treatment resistant Raynaud’s.

Methods: This new technique entails only a single-port endoscopic procedure, during which a sympathicotomy (figure 1) of the nerve is performed, thus sparing the ganglia. Hospital stay is limited to one day. The procedure has been developed for treatment of hyperhidrosis and performed in our hospital in over 550 patients without major adverse events with a success rate of 98%. In the current study, we aim to include 10 patients with treatment resistant Raynaud’s, defined as unsatisfactory effect or contraindications of oral vasodilatory agents and iv prostaglandin analogous. Sympathectomy was performed on the left hand first and the effects were compared with the contralateral hand after 3 and 12 months. Major exclusion criteria were severe lung involvement or proximal vascular stenosis. The primary end point was Raynaud’s Condition Score (RCS) and Quality of life (SF-36). Among others, cooling fingertip plethysmography (PPG) and laser doppler imaging (LDI) were used as secondary end points for objective assessment of hand perfusion.

Results: This is an interim report, and 4 patients have been included so far (age 20,29,32,56 years), male/female 3/1, 3 primary, 1 secondary), without a history of digital ulcers. No adverse events have been observed, apart from some hyperae mia in the first post-operative week. RCS significantly in each patient. Additionally, a clear improvement in hand perfusion following cooling was observed with PPG and LDI, as compared to the contralateral site. All 4 patients were satisfied and agreed to have their contralateral site operated on also. Patients will be followed in the outpatient clinic to assess long term efficacy.

Conclusions: Single-port thoracoscopic sympathicotomy is a novel minimally invasive technique which appears to be safe and feasible in patients with treatment resistant Raynaud’s phenomenon. However, this study is ongoing and long-term efficacy needs to be established.

Disclosure of Interest: None declared


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ASSESSING MORTALITY MODELS IN SYSTEMIC SCLEROSIS RELATED INTERSTITIAL LUNG DISEASE

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Background: Intestinal lung disease (ILD) is a major cause of morbidity and mortality in systemic sclerosis (SSc). However, the severity of lung involvement can vary widely, and current evidence-based treatment options have modest benefit. Prognosis assessment is particularly important for initial management decisions at the time SSc-ILD is diagnosed. There are now multiple mortality models available for use in SSc-ILD which utilise patient’s baseline parameters.

Objectives: The Gender, Age, and Lung Physiology (GAP) model,1 interstitial lung diseases –GAP (ILD-GAP) model,2 and the Smoking history, Age, and Diffusion capacity of the Lung (SADL) model3 were compared using a systemic sclerosis-ILD (SSc-ILD) cohort to evaluate which best determined prognosis.

Methods: The models were applied to a cohort of patients with SSc (meeting the 2013 ACR/EULAR classification criteria) seen at a tertiary care centre within 1 year of ILD diagnosis from 2000–2013. Demographics, clinical characteristics,