FRI0453

HOSPITALISATION AND SURVIVAL ANALYSIS IN SYSTEMIC SCLEROSIS PATIENTS WITH CONCOMITANT OR ISOLATED PULMONARY HYPERTENSION AND INTERSTITIAL LUNG DISEASE IN THE MULTIETHNIC SCLERODERMA COHORT SINGAPORE

M. Noviani[1], E.S. Seyed [2], S.M.Y. Kua [1], G.Y.L. Chan[3], G.G. Teng[4], W.G. Law[3], A. Santosa[4], A. Lim[4], J.L. Tan[5], E.L.W. Tay[6], J.W.L. Yip[6], S.C. Ng[1], A.H.L. Low[1]. *[1]Department of Rheumatology and Immunology, Singapore General Hospital; [2]Center for Quantitative Medicine, Duke-NUS Medical School; [3]Department of Rheumatology, Allergy and Immunology, Tan Tock Seng Hospital; [4]Department of Rheumatology, National University Health System; [5]Department of Cardiology, National Heart Center Singapore; [6]Department of Cardiology, National University Heart Center, Singapore, Singapore*

Background: Concomitant pulmonary hypertension and interstitial lung disease in systemic sclerosis (SSc-PH-ILD) represents a distinct subpopulation of SSc with poorer prognosis in Western studies. In Asian patients, characterisation of SSc-PH-ILD is still lacking.
Objectives: To analyse hospital admissions, survival and prognostic markers among SSc patients with PH, ILD or concomitant PH-ILD in the Scleroderma Cohort Singapore.
Methods: In this study involving 3 tertiary Rheumatology institutions Jan 2008 to , Oct 2016 SSc patients with significant pulmonary involvement were included. ILD was based on high resolution computed tomography and predicted FVC <70%. PH was based on either echocardiographic systolic pulmonary arterial pressure (sPAP) ≥50 mmHg, or right heart catheterization (RHC) findings of mean PAP≥25 mmHg. Hospitalisation rates and survival of SSc patients with PH, ILD or PH-ILD were compared. Risk factors of poor outcomes were identified by multivariate stepwise Cox regression analysis.
Results: Among 490 patients, 92 had ILD, 50 PH and 43 PH-ILD (table 1). Of 93 patients with PH or PH-ILD, 56 were based on echocardiography and 37 on RHC. The 5 year survival was 79%, 87% and 90% in PH, PH-ILD and ILD subgroup, respectively (figure 1). In multivariable analysis, PH was significantly associated with 2.8-fold increased risk of death. Male gender, malabsorption, digital ulcerations and renal crisis were also significantly associated with mortality (table 2). No significant difference in hospital admissions/year among different subgroups. Increased hospital admissions were associated with renal crisis, right heart failure and use of PH medications.

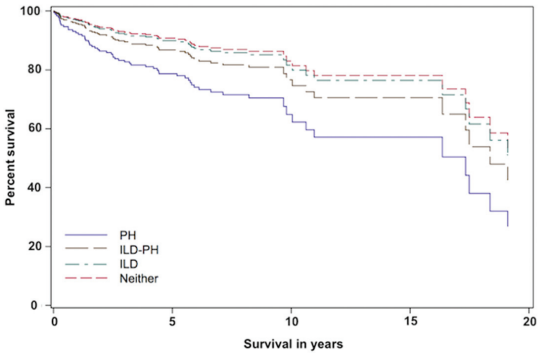
Abstract FRI0453 – Table 1. Clinical characteristics

	PH (n=50)	ILD (n=92)	PH-ILD (n=43)	No PH/ILD (n=305)
Female, n	44	76	38	270
Follow up duration (months ±SD)	53.46 ±55.76	101.5 ±80.04	88.71±65.53	64.66±36.27
Age at SSc diagnosis (years ±SD)	51.08 ±16.44	46.87 ±12.4	53.84±15.17	46.44±14.59
Duration of SSc at entry (years ±SD)	5.85±6.95	6.93±7.45	6.14±7.98	5.20±8.24
Dc-SSc, n	13	45	11	100
PH specific treatments[+], n	28	N/A	26	N/A
Immunosuppressants[++], n	25	64	27	176

[+]Prostacyclin, phosphodiesterase type 5 inhibitors, endothelin receptor antagonist; [++]Methotrexate, cyclophosphamide, mycophenolate mofetil.

Abstract FRI0453 – Table 2. Survival analysis

	Hazard Ratio (95% CI)	P Value
Male gender	2.85 (1.53–5.33)	0.0010
Malabsorption	2.89 (1.67–5.01)	0.0002
Renal crisis	2.00 (1.00–3.99)	0.0490
Digital ulcerations	2.06 (1.21–3.50)	0.0076



Abstract FRI0453 – Figure 1. Adjusted survival curve comparing survival of SSc patients with PH, ILD, and concomittant PH-ILD. X-axis shows years of survival from diagnosis of PH or ILD.

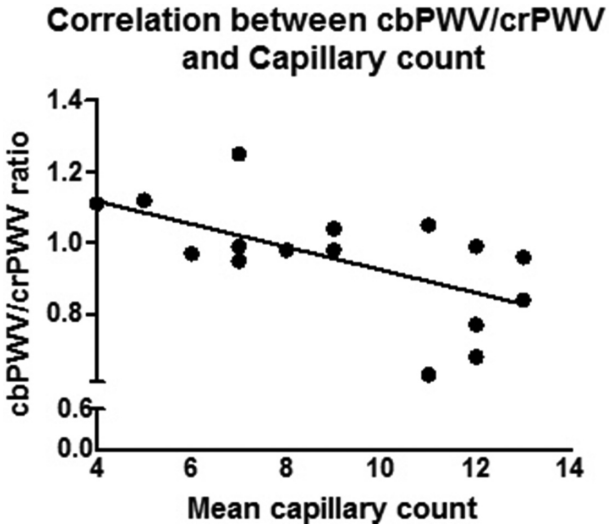
Conclusions: Compared to those with ILD or PH-ILD, SSc-PH patients had increased mortality, but not hospitalisation rates. This could be due to small sample size or short follow up duration. We identified risk factors associated with worse outcomes in SSc patients with significant pulmonary involvement.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.3185

FRI0454

ARTERIAL STIFFNESS OF THE FOREARM IS ASSOCIATED WITH NAIL-FOLD CAPILLARY COUNT IN SSC: A NOVEL MARKER OF EARLY VASCULOPATHY?

M.M. Habing[1], A. Eman Abdulle[1], A.M. van Roon[1], A.M. van Roon[1], H. van Goor[2], H. Bootsma[3], A.J. Smit[1], D.J. Mulder[1]. *[1]Dept. of Internal medicine, div. Vascular Medicine; [2]Dept. of Pathology and Medical Biology, Section Pathology; [3]Dept. of Rheumatology and Clinical Immunology, University of Groningen – University Medical Centre Groningen., Groningen, Netherlands*

Background: Microvascular disease, characterised by rarefaction of capillaries, is the hallmark of systemic sclerosis. Remarkably, obliteration of the ulnar and radial artery is regularly observed, implicating involvement of the larger forearm arteries. Pulse wave velocity (PWV) is a widely accepted non-invasive measure for arterial stiffness and may serve as an early biomarker of forearm artery involvement, before the occurrence of irreversible arterial obliteration.
Objectives: The aim of the current study was to investigate arterial stiffness of the aorta and of the upper extremities in SSc patients compared to healthy controls and to correlate these findings with nail-fold capillary count, skin involvement, and extent of disease.
Methods: In total, 19 SSc patients (median age 51 years, 68% female) and 19 age and gender matched healthy controls (median age 53 years, 68% female) were included. Patients characteristics were obtained and blood was drawn. Measurements of arterial stiffness were carried out by using the SphygmoCor System (AtCor Medical, Sydney, Australia) and pressure waveforms were measured at four sites, i.e. carotid, femoral, brachial, and radial. Aortic PWV was defined as carotid-femoral (cf) PWV. Upper extremity PWV was measured as carotid-brachial (cb) and carotid-radial (cr) PWV, and the ratio between cbPWV/crPWV was used as an indication of the relative change in PWV in the forearm. Capillary count was defined as the mean capillary count per 3 mm of 8 fingers. Skin involvement was assessed by the modified rodnan skin score. The number of ACR/EULAR 2013 criteria points was used as a surrogate for extent of disease.
Results: Upper extremity PWV measures were significantly higher compared to aortic PWV in patients and in controls (SSc: p<0.001; HC: p=0.03), but did not significantly differ between both groups (table 1). CbPWV/crPWV ratio correlated strongly with capillary count (r=−0.55, p=0.022, figure 1) in SSc patients with a borderline significant trend in regards to its relation with the extent of disease (r=0.48, p=0.053) and skin involvement (r=0.41, p=0.10).



Conclusions: Our findings demonstrate that arterial stiffness of the forearm has a relationship with nailfold capillary count and tends to be associated with the extent of disease in patients with SSc. These preliminary data may suggest that vascular damage may concomitantly occur in both capillaries as well as larger arteries of the forearm, which may potentially serve as a novel tool for assessing of early vascular involvement in SSc.

Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.7213

FRI0455 **INCREASING INCIDENCE OF ADULT IDIOPATHIC INFLAMMATORY MYOPATHIES: A TEN-YEAR UK EPIDEMIOLOGICAL STUDY**

M.J.S. Parker[1,2], A. Oldroyd[3], M.E. Roberts[4], R.G. Cooper[5], H. Chinoy[3], on behalf of UKMYONET. *[1]Rheumatology Department, Royal Prince Alfred Hospital, Sydney, Australia; [2]Rheumatology Department, Salford Royal NHS Foundation Trust, Salford; [3]Centre for Musculoskeletal Research, University of Manchester, Manchester; [4]Greater Manchester Neurosciences Centre, Salford Royal NHS Foundation Trust, Salford; [5]MRC-ARUK Centre for Integrated Research into Musculoskeletal Ageing, University of Liverpool, Liverpool, UK*

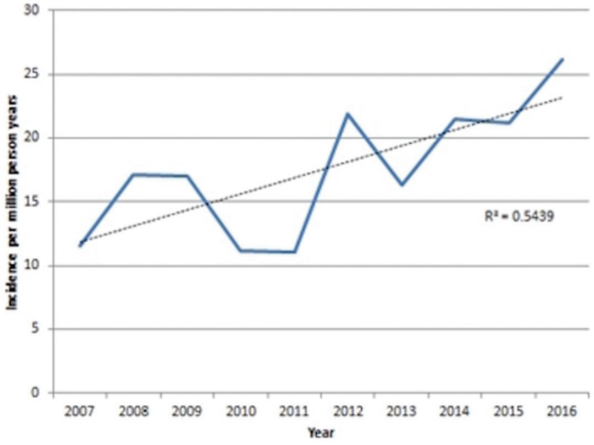
Background: Studying the epidemiology of rare conditions such as the idiopathic inflammatory myopathies (IIM) can assist in the identification of risk factors, disease associations and temporal trends. Interrogation of differing geographically and genetically diverse populations can help to construct a more complete picture of underlying disease patterns. A number of UK centres have contributed to national and international IIM research collaborations, but to date there has been no published report detailing the incidence or prevalence of adult IIM in the UK, or to establish the relative proportion of the varying clinical subtypes. Moreover, previous international studies have focussed on specific IIM subtypes, such as inclusion body myositis (IBM) or immune-mediated necrotising myopathy (IMNM), are historic, were undertaken before recent developments in our understanding of the range of IIM subtypes, and utilised widely varying methodologies and case acquisition strategies.

The recently published combined European League Against Rheumatism and American College of Rheumatology (EULAR/ACR) classification criteria for adult and juvenile IIM represent potential progress in identifying IIM, as well as various disease subtypes[1]. We present here the first epidemiological study to utilise these new criteria as part of disease verification.

Objectives: Identify and characterise all incident adult cases of IIM between Jan 1 st 2007 and Dec 31 st 2016 in the City of Salford, UK.

Methods: Adults first diagnosed with IIM within the study period were identified by: i) a Salford Royal NHS Foundation Trust (SRFT)inpatient episode IIM-specific ICD-10 coding search; ii) all new patient appointments to SRFT neuromuscular outpatient clinics; iii) all Salford residents enrolled within the UKMYONET study. All patients with 'definite' IIM by the 2017 EULAR/ACR classification criteria were included, as were 'probable' cases if expert opinion agreed. Cases were excluded if <18 years at disease onset, if they did not meet 'probable' criteria, or when 'probable' but expert opinion concluded a non-IIM diagnosis.

Results: The case ascertainment procedures identified 1156 cases which, after review and application of exclusion criteria, resulted in 32 incident cases during the study period. 23/32 were female with a mean age of 58.1 years. The mean incidence of adult IIM was 17.6/1,000,000 person years (py), higher for females than for males (25.2 *versus* 10.0/1,000,000py respectively). A significant incidence increase over time was apparent (13.6 *versus* 21.4/1,000,000py; p=0.032). Using EULAR/ACR classification criteria, the largest IIM subtype (21/32) was polymyositis, followed by dermatomyositis (8/32), inclusion body myositis (2/32) and amyopathic dermatomyositis (1/32). Expert opinion subtype differed from EULAR/ACR Classification criteria in 19/32 cases.



Conclusions: The incidence of adult IIM in Salford is 17.6/1,000,000py, higher in females and is increasing over time. Disagreement exists between EULAR/ACR-derived and expert opinion-derived IIM subtype assignments.

REFERENCES:

[1] Lundberg IE, Tjärnlund A, Bottai M, et al. EULAR/ACR Classification Criteria for Adult and Juvenile Idiopathic Inflammatory Myopathies and their Major Subgroups. Ann Rheum Dis. 2017;76:1955–64.

Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.6079

FRI0456 **PREDICTORS FOR DISEASE WORSENING DEFINED BY ORGAN FAILURE IN DIFFUSE SYSTEMIC SCLEROSIS: A EUROPEAN SCLERODERMA TRIALS AND RESEARCH (EUSTAR) ANALYSIS**

M.O. Becker[1], N. Graf[2], R. Sauter[3], Y. Allanore[4], J. Curram[5], C. Denton[6], D. Khanna[7], M. Matucci-Cerinic[8], J. Pena[9], J.E. Pope[10], O. Distler[1]. *[1]Department of Rheumatology and the Centre of Experimental Rheumatology, University Hospital Zurich, Zurich, Switzerland; [2]Graf Biostatistics, Winterthur, Switzerland; [3]Big Data Institute, Li Ka Shing Centre for Health Information and Discovery, Nuffield Department of Medicine, University of Oxford, Oxford, UK; [4]Paris Descartes University, Sorbonne Paris Cité, Rheumatology A Department, Cochin Hospital, Paris, France; [5]Bayer Plc, Reading, UK; [6]UCL Division of Medicine, Royal Free Campus, London, UK; [7]Division of Rheumatology, Department of Internal Medicine, University of Michigan Scleroderma Program, University of Michigan, Ann Arbor, MI, USA; [8]Department of Experimental and Clinical Medicine, University of Florence, Florance, Italy; [9]Bayer US LLC, Whippany, NJ, USA; [10]Department of Medicine, Division of Rheumatology, University of Western Ontario, St. Joseph's Health Care, London, ON, Canada*

Background: Mortality and worsening of organ function are desirable endpoints for clinical trials in systemic sclerosis (SSc). However, these events are relatively rare, making clinical trial design challenging.

Objectives: To identify factors in a population of patients with diffuse SSc from the European Scleroderma Trials and Research (EUSTAR) group database that predict these endpoints and hence allow enrichment of those patients.

Methods: Inclusion criteria were a diagnosis of diffuse SSc and follow-up after 9–15 (12±3) months. This timeframe was chosen to reflect typical clinical trial design. Disease worsening/organ progression was fulfilled if any of the following events occurred: new renal crisis, decrease in forced vital capacity (FVC) ≥10%, new left ventricular ejection fraction (LVEF) <45% or decrease in LVEF by >10% for patients with baseline LVEF <45%, new pulmonary (arterial) hypertension, or