Conclusion: Patients with systemic sclerosis positive for anti-U1RNP differ in the predominance of inflammatory musculoskeletal manifestations and frequent combination with Sjogren's syndrome and overlaps. Highly positivity for anti-U1RNP is accompanied by a persistent increase in RF, anti-Ro, anti-dsDNA

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AB0610   **SEASONAL VARIATION IN IDIOPATHIC INFLAMMATORY MYOPATHIES INCIDENCE AND PRESENTATION: A RETROSPECTIVE STUDY IN BEIJING AND HONG KONG**

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Background: Seasonal patterns of disease onset and severity in idiopathic inflammatory myopathies (IIMs) as a whole are conflicting [1-3]. In recent years, over 10 myositis-specific antibodies (MSAs) have been identified. They are able to divide patients into homogenous subgroups and inform on prognosis [4].

Objectives: The objective of the study was to investigate the seasonal variation of onset of IIMs characterised serologically.

Methods: This was a multi-centred retrospective observational study. Consecutive Chinese patients with IIMs admitted to the rheumatology wards of the participating major regional hospitals in Beijing and Hong Kong from July 2013 to June 2018 were recruited. The diagnosis of IIMs was based on the Bohan and Peter's criteria with definite or probable cases being included [5]. Patients with clinically amyopathic disease must have the typical Gottron's papules or heliotrope rash as determined by rheumatologists or dermatologists, and with no symptoms or signs of muscle involvement according to Sontheimer [6]. Patients with juvenile myositis, inclusion body myositis, cancer-associated myositis and myositis associated with other connective tissue disease were excluded. A commercial line blot immunoassay kit (EUROLINE) was used to detect the MSAs.

Results: All together 495 patients were studied. The mean age of the patients at disease onset was 48.1 years (S.D. 13.3). There was a female predominance (68.3%). The subgroups of IIMs were: dermatomyositis (61.0%), polymyositis (21.8%), clinically amyopathic dermatomyositis (12.9%), immune mediated necrotising myopathy (3.8%) and nonspecific myositis (0.4%). No particular seasonal pattern in disease onset was observed in IIM patients as a whole (Figure 1) or in any classical subgroups. However, significantly more patients with any one MSA had their disease started in the first half of the year (p=0.007) as shown in Figure 2. Patients with either anti-synthetase or anti-MDA5 antibodies, which are associated with interstitial lung disease, had more frequent disease onset from November to February, which might coincide with the local flu season. It was also found that MSA positivity was associated with infection of the patient (p=0.005). Further analyses showed that patients with MSAs which are typically associated with severe skin disease (MDA5, TIF1g, NXP2, SAE) had more hospitalisation from April to September where excessive sun exposure is expected. There were no major differences between the Beijing and Hong Kong subgroups.

Conclusion: Apparent seasonal patterns were noticed in our ethno-serologically defined IIM patients. Certain environmental factors, particularly infection or UV exposure, could be potential triggers. Our findings could shed light on the identification of etiologic factors and enhance our understanding of disease pathogenesis.

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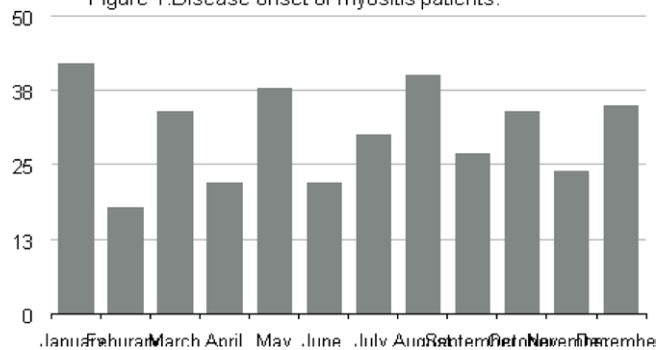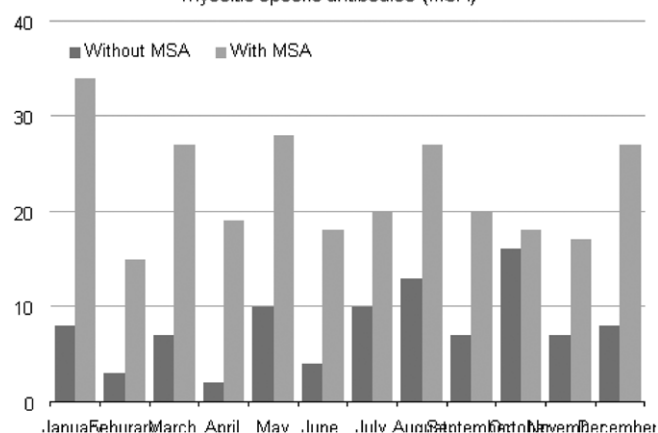
Figure 1. Disease onset of myositis patients.

Figure 2. Disease onset of myositis patients with or without myositis specfic antibodies (MSA)

Disclosure of Interests: None declared

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AB0611   **STRAIN ANALYSIS OF THE RIGHT VENTRICLE USING 2D-SPECKLE TRACKING ECHOCARDIOGRAPHY IN A COHORT OF PATIENTS WITH SYSTEMIC SCLEROSIS**

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Background: Systemic Sclerosis (SSc) is a rare and life-threatening connective tissue disease with multiple organ impairment. Cardio-pulmonary involvement is common: pulmonary fibrosis, pulmonary hypertension (PH), and electrical disorders are the most serious complications and causes of increased mortality.

Objectives: We evaluated features related with the onset and development of PH in a cohort of SSc patients. We further studied ecocardiographic abnormalities, by means of 2D-speckle tracking echocardiography (STE) with specific reference to the right ventricular strain measure (RV-strain).

Methods: We analyzed data from 50 SSc patients (pts) referred to our University-based Rheumatology Centre and SSc Unit from January 2007 to June 2019 (F/M 45/5; lc/dcSSc 45/5; mean age 59.20±14.357 years; mean disease duration 12.08±8.75 years). All pts underwent general and cardio-pulmonary assessment in our Cardio-Rheumatology Clinic. The following parameters were considered: blood exams, in particular inflammation indexes, uric acid test and serum autantibodies; pulmonary function tests; high resolution scan of the lungs (HRCT); standard electrocardiogram (ECG) and RV-strain measured by 2D-STE. These examinations were performed according to clinical picture and current methodologies. We compared SSc subjects with (10/50) and without (40/50) PH diagnosis during follow-up regardless of treatments.

Results: SSc pts with PH didn't show significant alterations concerning RV-strain if compared with pts without PH (p=0.707). Nevertheless, RV-strain value was modified in relation to TAPSE alterations in all pts but this data correlated with right ventricular dilatation only in PH subjects. Furthermore, interesting significant values about dilatation of right and left atria (p=0.007, p=0.048), dilatation of inferior vena cava (p=0.037) and right ventricle (p=0.023) were observed. Left ventricular hypertrophy (p=0.012) as well as valvular insufficiencies (mitral and aortic) were more frequent in PH group too (p=0.016). These pts showed higher incidence of

skin ulcers (p=0.0001), higher values of blood pressure (p=0.004), elevated uric acid levels (p=0,027) and anti-centromere antibodies positivity (p=0.0001).

Conclusion: Our research provides further evidence of the prognostic value of echocardiographic findings in SSc subjects, with focus on PH. Population enlargement is ongoing in order to identify more accurate results about RV-strain, considering the efficacy of PH treatments on cardiac contractility. Speckle tracking echocardiography proves to be a sensitive, low-cost, non-invasive and reliable tool to detect early cardiac impairment in Ssc, full of potential future prospects.

Disclosure of Interests: None declared

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SHORT-TERM REVERSIBLE IMPROVEMENT IN EARLY-PHASE ELEMENTS OF NAILFOLD CAPILLARY ABNORMALITIES IN PATIENTS WITH SYSTEMIC SCLEROSIS BY INTRAVENOUS CYCLOPHOSPHAMIDE (IVCY)

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Background: Nailfold capillary abnormalities are one of representative signs in systemic sclerosis (SSc). However, previous reports about changes in nailfold capillary by immunosuppressive therapy have been limited. Especially, there have been no reports about short-term changes in nailfold capillary abnormalities.

Objectives: To clarify whether intravenous cyclophosphamide (IVCY) treatment for SSc patients can improve nailfold capillary abnormalities in half a year.

Methods: Among patients diagnosed as having SSc according to the 2013 ACR/EULAR classification criteria at our hospital from May 2018 to December 2019, those who treated with IVCY for interstitial lung disease (ILD) were consecutively registered. All patients received IVCY six times. Nailfold capillary abnormalities on eight fingers including both second to the fifth fingers were observed with a nailfold videocapillaroscopy (NVC). Each finger was evaluated for enlarged capillary, giant capillaries, hemorrhage, loss of capillary, disorganization of the vascular array, and capillary ramification. Quantitative scoring was performed on a scale of 0 to 3 in accordance with the ratio of each of them. NVC tests were evaluated before IVCY treatment intervention and after IVCY. In all cases, the evaluation of NVC after IVCY treatment was performed 6 months after the administration day. Skin changes were evaluated by modified Rodnan's total skin thickness score (mRSS) at performing NVC. Anti-centromere antibodies, anti-Scl-70 antibodies, anti-RNA polymerase III, and anti-RNP antibodies were measured. Pulmonary function tests (PFTs) including forced vital capacity (FVC) and diffusing capacity of the lung carbon monoxide (DL_{CO}) were performed before and after IVCY. The statistical significance of the differences between means of two groups was evaluated by paired t-test. A p level of 0.05 or less was considered statistically significant.

Results: Five patients were included. The mean age was 59 years and 4 patients were female (80%). High dose corticosteroids were used in 2 patients (40%). Anti-RNA polymerase III was positive in 2 patients (40%), anti-Scl-70 antibody was positive in 1 (20%), and negative test for any specific antibodies was in 2 (40%). Changes in NVC scores, which were total scores of 8 fingers, were as follows: Enlarged; 13.2±4.8 to 6.4±5.9 (p=0.018), Giant; 7.0±5.7 to 1.6±1.1 (p=0.0314), Hemorrhage; 8.4±6.2 to 3.2±2.3 (p=0.0274), Loss; 4.0±2.5 to 0.6±1.3 (p=0.0288), Disorganization; 0.6±0.9 to 1.0±1.0 (p=0.7065), Ramification; 0.6±0.9 to 0.8±1.8 (p=0.5730). (Table) After IVCY treatment, mRSS reduced in 4 cases (80%). Changes in mRSS scores were as follows: 18.8±8.3 to 12.4±13.3 (p=0.0677). The cases with improved mRSS and those with improved NVC findings were consistent. The mean FVC before and after IVCY was 2077 ml and 2062 ml, respectively. The mean DL_{CO} before and after IVCY was 9.88 mL/min/mmHg and 9.58 mL/min/mmHg, respectively.

Conclusion: Nailfold capillary abnormalities in patients with SSc could be improved in half a year with IVCY. Especially, early phase elements including enlargement, giant, and hemorrhage were specifically reversible.

Table.

No.	(E)	(G)	(H)	(L)	(D)	(R)	mRSS
1	21→14	16→3	19→7	0→0	0→2	0→0	14→9
2	12→3	4→2	6→1	4→0	2→1	1→0	15→1
3	14→11	8→2	6→2	7→0	0→0	0→4	10→5
4	10→4	6→1	8→3	4→3	0→2	2→0	25→12
5	9→0	1→0	3→3	5→0	1→0	0→0	30→35
mean ± SD	13.2±4.8	7.0±5.7	8.4±6.2	4.0±2.5	0.6±0.9	0.6±0.9	18.8±8.3
	6.4±5.9	1.6±1.1	3.2±2.3	0.6±1.3	1.0±1.0	0.8±1.8	12.4±13.3
p-value	0.018	0.0314	0.0274	0.0288	0.7065	0.5730	0.0677

E: enlarged, G: giant, H: hemorrhage, L: loss, D: disorganization, R: ramification.
The table shows the total of eight points for each finding in the NVC test. The previously described values are before treatment and the later values are after treatment.

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AUTONOMIC NEUROPATHY AND ITS PREDICTORS IN SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc), a chronic autoimmune disease, is associated with autonomic neuropathy[1]. Autonomic neuropathy, especially cardiovascular autonomic neuropathy (CAN) is significant risk predictor of sudden cardiac death. However, its relationship with disease specific measures remains unexplored in SSc.

Objectives: To assess cardiovascular autonomic neuropathy and sudomotor function and its predictors in systemic sclerosis.

Methods: In this cross-sectional study, 16 SSc patients meeting the 2013 European League Against Rheumatism (EULAR) and American College of Rheumatology (ACR) classification criteria and 15 age and sex-matched healthy controls were recruited. Cardiovascular autonomic function assessed by five cardiovascular reflex tests according to Ewing. Peripheral sympathetic autonomic function assessed by FDA approved Sudoscan (Impeto Medical, Paris) through measurement of electrochemical skin conductance. Disease-specific measures (Disease duration, Modified Rodnan Skin Score (mRSS), EUSTAR activity score), and inflammatory measures (ESR, CRP) were determined. Quality of life measured by Scleroderma Health Assessment Questionnaire (SHAQ).

Results: Systemic sclerosis patients had significantly impaired parasympathetic [Heart rate response to deep breath (HRD) (Fig. 1A), Heart rate response to standing (HRS) (Fig. 1B) and Heart rate response to valsalva manoeuvre (Fig. 1C)] and symapathetic [BP response to hand grip (BPH) (Fig. 1D)] function as compared to healthy controls. Scleroderma patients had significantly impaired sudomotor function (p<0.05) as compared to healthy controls. Levels of mRSS, EUSTAR score, ESR, CRP and SHAQ were significantly higher in SSc patients as compared to healthy controls (p≤0.05). Parasympathetic (HRD & HRS) dysfunction inversely correlated with ESR, CRP and mRSS. Sudomotor function positively correlated with mRSS, disease duration and CRP.

Conclusion: CAN and Sudomotor function are significantly impaired in SSc. Parasympathetic dysfunction is more pronounced than sympathetic dysfunction in SSc. CAN and Sudomotor dysfunction are associated with disease-duration, skin-score, ESR and CRP. These could serve as potential predictors of Cardiovascular Autonomic neuropathy and sudomotor dysfunction in SSc.

Fig 1A. Significant difference in HRD between SSc and healthy controls (*p≤0.05)

Fig 1B. Significant difference in HRS between SSc and healthy controls (p≤0.05)

Fig 1C. Significant difference in HR response to valsalva manoeuver between SSc and healthy controls (p≤0.05)

Fig 1D. Significant difference between BP response to hand grip as compared to healthy controls (p≤0.05)