CTD among the enrolled patients (61%), followed by MCTD (16%). Hemodynamic characteristics and findings after exercise echocardiography in patients having normal mean PAP, borderline mean PAP, and PH were compared. We also determined the validity of therapeutic intervention.

Results: The values of tricuspid regurgitation pressure gradient (TRPG) were comparable between patients with borderline (31.1±7.8 mmHg) and normal mean PAP (28.3±6.9 mmHg) (P=0.1572) but its value became significantly higher in patients with borderline mean PAP (39.1±8.0 mmHg) than in those with normal mean PAP (32.8±7.4 mmHg) after exercise echocardiography (P=0.0391). Pulmonary arterial wedge pressure was significantly elevated in patients with borderline mean PAP (12±3 mmHg) compared with that in normal mean PAP (7±3 mmHg) ($P<0.0001$) and its value was comparable to those with overt PH, suggesting the heterogeneity on the cause of borderline mean PAP among CTD patients. The clinical course of 10 patients with borderline mean PAP was studied. Five were treated for precapillary disease, 3 for postcapillary disease, and 2 for interstitial lung disease (ILD). Normalization of mean PAP was seen in 3/4 and 3/3 of the patients treated for precapillary and postcapillary disease, respectively. Deterioration of TRPG was seen in one patient after receiving pulmonary vasodilators. One with severe ILD developed PH.

Conclusions: The pathogenesis of borderline mean PAP, clearly distinctive from normal mean PAP, was heterogeneous as that of manifest PH in CTD patients. Though the clinical course may be altered with appropriate therapeutic intervention, repeated assessment is needed.

References:

[1] Kovacs G, Olschewski H. Borderline pulmonary pressures in scleraderma - "a pre-pulmonary arterial hypertension" condition? Arthritis Res Ther 2015; 17: 123, 015–0649–7.

[2] Kovacs G, Avian A, Tscherner M, et al. Characterization of patients with borderine pulmonary arterial pressure. Chest 2014; 146: 1486–93.

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SAT0370 EVALUATIONS OF EXERTION DYSPNEA IN PATIENTS WITH CONNECTIVE TISSUE DISEASE (CTD) BY CPET (CARDIOPULMONARY EXERCISE TESTING) FOR EARLY DETECTING ASSOCIATED PULMONARY HYPERTENSION (APAH)

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Background: Patients with CTD often complain of exertion dyspnea, due to lung diseases, heart diseases, musculoskeletal disorders and/or APAH. Although imaging and physiological tests could reveal pathophysiology, some of them remain unknown. Especially APAH is rarely diagnosed in WHO functional class I or II, because pulmonary vasculopathy (PV) cannot be detected until two-thirds of pulmonary blood vessels deteriorated, [1] with cardiac ultrasonography (UCG) or right heart catheterization (RHC) at rest. Pathophysiological considerations suggest that the haemodynamic/metabolic impairment in APAH may be observed during exercise before the disease becomes evident at rest.[2–3] We evaluated exertion dyspnea of unknown to find out early APAH with CPET.

Objectives: We performed CPET in patients who complained of exertion dyspnea and tried to detect early PV.

Methods: From June 13th in 2015 to October 28th in 2016, we performed CPET and evaluate their clinical state in 28 patients, 17–80 years 2 males/26 females, 5 mixed connective tissue disease (MCTD), 15 SSc, 3 SLE, 2 Sjögren's syndrome, 3 dermatomyositis. They underwent UCG, pulmonary function testing (PFT), 6-minutes walk test, and nailfold capillaroscopy by Cutolo's method.[4]

Results: Twenty cases presented decreased peak VO2. VE/VCO_2 ratio, which represent increased ventilation-perfusion mismatch, elevated in 12 cases. 8 cases, with decreased peak VO2 but normal VE/VCO_2, were regarded that muscle weakness mainly induced exertion dyspnea, and advised exercise. Twelve cases with decreased peak VO2 and elevated VE/VCO_2 were estimated to have APAH or/and interstitial lung disease (ILD). Seven of them underwent RHC and 1 case diagnosed as definite APAH, and another 1 case as post capillary PH.

A 34 years MCTD woman without ILD showed an active capillaroscopic pattern, her peak VO2 decreased (13.9ml/kg/min.) and nadir VE/VCO_2 elevated (39). Although her mean PAP was normal, we suspected she had early PV and administered PDE5 inhibitor to her and her dyspnea had gone soon and CPET parameters improved.

CPET was also useful for early detections of therapeutic gains in APAH. A SSc woman, diagnosed as APAH by RHC, was performed CPET only 9 days after administration of PDE5 inhibitor. Her peak VO2 elavated (13.3 to 15.4 ml/kg/min.) and nadir VE/VCO_2 decreased (43.4 to 38.9) promptly.

Thirteen patients showed an early capillaroscopic pattern, 7 an active pattern and 3 a late pattern. Median values of minimum VE/VCO_2 ratio significantly differed ($p<0.05$) in the three capillaroscopic groups, being progressively worse from early to late capillaroscopic pattern.

Conclusions: We performed CPET in every patient safely. Although more research is required, CPET may provide valuable information notably in APAH patients.

References:

[1] Hachulla E, et al. French PAH-SSc Network. PAH with SSc in patients with functional class? dyspnea; mild symptoms but severe outcome. Rheumatology 2010;49:940–4.

[2] Grunig E, et al. Non-invasive diagnosis of pulmonary hypertension: ESC/ERS Guidelines with Updated Commentary of the Cologne Consensus Conference 2011. Int J Cardiol 2011;154:53.

[3] Domitrescu C, et al. Developing PV in SSc, detected with non-invasive CPET. PLoS ONE 2010;5e14293.

[4] Cutolo M, et al. Nailfold videocapillaroscopy assessment of microvascular damage in SSc. J Rheumatol. 2000 Jan;27(1):155–60.

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SAT0371 BONE FRACTURE RISK ASSESSMENT IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES: RESEMBLANCES TO RHEUMATOID ARTHRITIS

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Background: Idiopathic inflammatory myopathies are chronic, heterogeneous systemic autoimmune diseases with symmetrical proximal muscle weakness. During the disease course, osteoporosis and bone fractures are more common compared to the healthy population, which can be explained by the chronic inflammation, immobilization, spontaneous falls and steroid treatment, and affect crucially the patients' quality of life. Recently, a WHO fracture risk calculation tool, FRAX score is available, to measure the 10-year probability of osteoporotic fractures. It takes into account relevant clinical risk factors, such as rheumatoid arthritis, however myositis does not exist among the risk factors.

Objectives: Estimation the effect of myositis and myositis related bone mineral densities on bone fracture risk calculated by FRAX tool.

Methods: FRAX score was determined in 71 patients with idiopathic inflammatory myopathies and results were compared with the data from 50 age, sex and BMI matched patients with rheumatoid arthritis. Moreover, osteoporosis related biomarkers, disease related fractures and bone mineral densities were determined using DXA examinations. Statistical analysis was performed with IBM SPSS 20.0 software.

Results: There were no significant differences between the demographical data, biomarkers (Ca, Vitamin D, parathormone level) of the two groups. Disease duration and cumulative steroid dose were higher in the myositis group. Results of the FRAX score without BMD were significantly lower in the patients with myositis, in both fracture risk: major osteoporotic (8,61±6,36%, vs. 15,59±12,66%; p: 0,002) and femur neck (2,66±3,24%, vs. 6,34±9,018; p: 0.003). T score results of the DXA examination were not significantly different between the two populations (Lumbar1–4: -0,9±1,43 vs.-0,829±1,38; p:0,829; Femoral neck: -1,4±1,08 vs. -1,02±1,08; p: 0,93), but the presence of osteopenia (60% vs. 39,5%) and osteoporosis (13,5% vs. 7%) were more frequent in the myositis group (p: 0,045). Disease related fracture was associated with disease duration in the myositis group and with antibody (RF, ACPA) presence in the RA group. FRAX score with BMD results showed no significant differences between the two populations (Major osteoporotic: 9,44±6,72 vs. 13,25±9,43; p: 0,053; Hip: 2,77±3,01 vs. 3,57±5,08; p: 0,811).

Conclusions: As far as we know, this is the first study which examine the fracture risk using FRAX score in patients with idiopathic inflammatory myopathies. According to our data, we can conclude that existence of myositis might indicate similar, independent risk factor in fracture probability, like rheumatoid arthritis. Evaluation of fracture risk should be done with DXA result in patients with IIM, otherwise risk could be underestimated. An exact value of the "myositis related risk" could be determined by a 10-year prospective study.

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SAT0372 CLINICAL AND SEROLOGICAL ASSOCIATIONS OF AUTOANTIBODIES TO BICD2 AS A NOVEL MARKER FOR SYSTEMIC SCLEROSIS

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Background: Anti-nuclear antibodies (ANA), which are present in approximately 90% of systemic sclerosis (SSc) patient's sera, play an important role in establishing the diagnosis and predicting prognosis of SSc. Recently, a novel autoantibody has been described in SSc patients targeting Cytoskeleton-Like Bicaudal D Protein Homolog 2 (BICD2).

Objectives: The aim of this study was to assess the prevalence and titers of anti-BICD2 antibodies in SSc and controls and to study the clinical associations of this new antibody.

Methods: A total of 502 samples from SSc patients enrolled in the Canadian Scleroderma Research Group (CSRG) cohort were included in this study. Clinical associations were assessed either as anti-BICD2 antibody positivity