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FRI0446 SEVERE HEART DISEASE IN SYSTEMIC SCLEROSIS: PREVALENCE, RISK FACTORS AND CURRENT TREATMENT. A EUSTAR-DESSCIPHER STUDY

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Background: Heart disease in patients with systemic sclerosis (SSc) can cause manifestations such as cardiac blocks (CBs), ventricular arrhythmias (VA), and congestive heart failure (CHF) which may require a pacemaker/defibrillator implant, are the cause of death or sudden death (SD) in about 20% of patients and can be referred to as severe heart disease (SHD). The European League against Rheumatism – Scleroderma Trial and Research (EUSTAR) database provides an unique opportunity to address this topic. The results of such analysis are instrumental for the DeSScipher project, an EU funded project devoted to *decipher the optimal management of SSc*.

Objectives: To investigate the prevalence of each SHD manifestation and their associations with demographic, serological, clinical and therapeutic aspects in a large series of adult SSc patients.

Methods: Seven EUSTAR-DeSScipher centers provided clinical charts at study entry including all the items of SHD. The prevalence of any and each SHD manifestation was calculated. The associations with demographic, serological, clinical features and treatment with vasodilators, i.e. calcium channel blockers (CCBs) and /or ACE inhibitors (ACEi), were investigated by univariate analysis and confirmed by multivariate logistic regression analysis.

Results: At July 6th 2012, 1119 SSc patients from the 7 EUSTAR centers had no missing data (995 females, 164 males; median age 53.6 years, range 14-86). Out of them, 211 patients (19%) had at least 1 SHD manifestation, 152 patients had CBs (14%), 71 had VA (6%), 132 had CHF (12%), and 27 received a pacemaker/defibrillator implant (2%). CBs were the only SHD manifestation in 100 patients, VA in 31, and CHF in 80. No association was found between SHD and clinical (diffuse or limited) and serological (ACA or anti-Scl-70) subset. No association was found between either CHF or pacemaker/defibrillator implant and any other disease feature. In multiple logistic regression analysis, CBs were associated with bibasilar lung fibrosis at chest X-Ray (OR=2.6; 95%CI=1-6.4; p=0.04), while VA were associated with increased CPK levels (OR=11; 95%CI=1-117; p=0.02) and, unexpectedly, current CCB use (OR 10; 95%CI 1.4-76.1; p=0.02).

Conclusions: This is the first study devoted to investigate SHD in a large series of carefully assessed SSc patients. SHD was detected in about 19% of the cases, CBs in 14%, VA in 6%, CHF in 12%. Pacemaker/defibrillator implant was needed in 2.4%. These data highlight the importance of an accurate heart assessment in SSc patients. Moreover, the associations between VA and CPK elevation and current CCB use should stimulate the clinician to avoid these drugs in patients with myositis/myocarditis.

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FRI0447 TEMPOROMANDIBULAR JOINT AND CHEWING FEATURES IN SYSTEMIC SCLEROSIS PATIENTS: A CLINICAL AND MAGNETIC RESONANCE IMAGING ANALYSIS

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Background: The pathogenesis of Systemic Sclerosis (SSc) remains unknown, but increasing evidence suggests that activation of lesional fibroblasts contributes to the fibrotic process. SSc is characterized by symmetric, erosive synovitis, which may result in joint irregularity and disability.

Objectives: The purpose of this clinical study was to assess the prevalence and characteristics of Temporomandibular joint (TMJ) symptoms, clinical and Magnetic Resonance Imaging (MRI) findings in a cohort of patients with SSc.

Methods: 27 patients with SSc (12 Diffuse, 15 Limited, mean age 53.9, SD ±1.2) and 28 healthy subjects (mean age 54.8, SD ±4.2) were enrolled in this cohort study. Oro-facial clinical examination for assessing the presence of TMJ sounds, pain in the TMJ area, tenderness of masticatory muscles, limited mouth opening, pain assessment, MRI scan and Anamnestic and Dysfunctional Index were achieved in all of patients.

Results: The test groups presented more clinical and MRI TMJ symptoms and dysfunction than Control group. The distributions of symptoms were significantly different ($P<0.05$), in the Test groups for TMJ sounds, pain during mandibular movement and difficulty in the maximum mouth opening. There was also a significant decrease ($P<0.001$), in the Test groups, in the mean of leftward, rightward laterotrusion and protrusion. Correlation analysis showed that maximum opening leftward laterotrusion, protrusion and click were significantly correlated to Modified Rodnan Skin Score. The mean duration of disease was significantly correlated, ever in Total SSc group, only for the maximum mouth opening value.

Conclusions: This study proves that TMJ dysfunction is a common feature in SSc patients and it is correlated with a extent and involvement of disease and supports the notion that TMJ examination should be encouraged in rheumatology and clinicians should provide a patient support and a right pain management.

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FRI0448 PAH RISK CALCULATOR (DETECT), BESIDES SSC-PAH, REVEALS SSC PATIENTS WITH BORDERLINE AND EXERCISE-INDUCED PH

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Background: Early diagnostics PAH, associated with SSc is very important, due to devastating nature of the disease. Borderline and exercise-induced PH is a condition thought to represent early-stage pulmonary arterial vasculopathy.

Methods: We selected 21 patients with SSc (age 52+10 years, all female) with normal resting mean PAP (17 (14;21) mm Hg) who underwent graded bicycle supine exercise right heart catheterization (RHC). Patients were included in this study if they had 1) a pulmonary artery wedge pressure (PAWP) $\leq$15 mmHg by RHC; 2) no significant interstitial lung disease; 3) no left heart diseases like systemic hypertension, ischemic heart disease etc. Mean pulmonary artery pressure (mPAP), cardiac output (CO), and others were estimated invasively. Cut-off values of mean PAP $\geq$30 mm Hg at peak exercise were considered exercise-induced pulmonary hypertension (EI-PH), mean PAP 21-24 mm Hg so-called borderline pulmonary hypertension (Bo-PH). The risk of PAH development was calculated by DETECT PAH risk calculator.

Results: 11 of 21 pts had risk factors by DETECT calculator. We didn't find PH at rest RHC in none of 21 pts. 6 SSc pts had mPAP level between 21-24 mm.Hg. 10 (48%) patients showed EI-PH (median level of mPAP =36 (32; 42) mm Hg), 7 hadn't changes in pressure follow exercises. 4 pts were excluded due to low exercise tolerance. We revealed significant correlation between the level of mPAP and DETECT total risk score ($r=0,55$, $p<0,01$). We found that patients with EI-PH