SAT0253 PROGNOSTIC VALUE OF CARDIAC MAGNETIC RESONANCE IN SYSTEMIC SCLEROSIS

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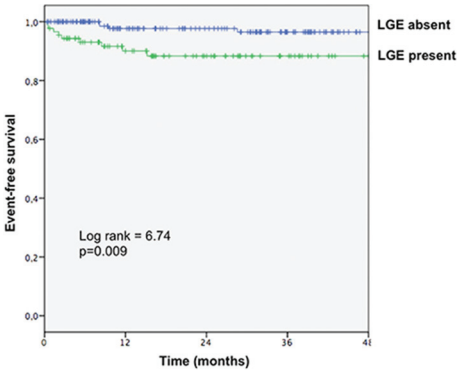
Background: Cardiac involvement is frequent in patients with systemic sclerosis (SSc). It is often subclinical, but significantly affects the prognosis of the disease. Cardiovascular magnetic resonance (CMR) is the non-invasive gold standard to quantify biventricular functional parameters and to perform a myocardial tissue characterization.
Objectives: To evaluate the prognostic value of CMR for cardiac events in SSc.
Methods: Two hundred and seventy-three SSc patients with a thorough clinical assessment underwent a CMR exam using a 1.5 T GE scanner. We quantified biventricular function parameter by SSFP cine images, oedema by STIR T2 images, and macroscopic fibrosis by late gadolinium enhancement (LGE) technique. Patients were followed-up and cardiac events were recorded as new onset of pulmonary arterial hypertension (PAH), new onset of heart failure (HF), or at least sustained ventricular tachycardia.
Results: Mean follow-up was 23.9 ± 17.0 months. During the follow-up a total of 14 events occurred (3 new onset PAH, 5 new onset HF, 6 ventricular tachycardia). CMR predictors of cardiac events by univariate analysis were left and right ventricular ejection fractions, indexed left and right atrial areas, and LGE (see Table). Myocardial fibrosis by LGE was the only independent predictor at multivariate analysis (hazard ratio 3.175; 95% C.I. 1.021-9.870, see Figure).

	Univariate analysis		Multivariate analysis	
	HR (95% CI)	p	HR (95% CI)	p
Left Ventricular Ejection Fraction	0.904 (0.843 – 0.969)	**0.004**	0.935 (0.868 – 1.007)	0.075
Indexed Left Atrial Area	1.244 (1.002 – 1.546)	**0.048**		
Right Ventricular Ejection Fraction	0.926 (0.884 – 0.969)	**0.001**	0.958 (0.904 – 1.014)	0.139
Indexed Right Atrial Area	1.384 (1.059 – 1.809)	**0.017**		
Right Ventricular End-Diastolic Mass	1.039 (0.991 – 1.091)	0.115		
Right Ventricular End-Systolic Mass	1.036 (0.988 – 1.086)	0.148		
T2 ratio	0.247 (0.019 – 3.251)	0.288		
Late Gadolinium Enhancement	3.871 (1.289 – 11.623)	**0.016**	3.175 (1.021-9.870)	**0.046**



Conclusion: Cardiac magnetic resonance anatomical and functional parameters of both the left and right heart have significant prognostic value in patients with SSc.
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SAT0254 VASODILATOR THERAPY IN THE LONG TERM PREVENTION OF MYOCARDIAL MANIFESTATIONS IN SYSTEMIC SCLEROSIS (SSC): RESULTS FROM DESSCIPHER INCEPTION COHORT STUDY

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Background: Calcium channel blockers (CCB) and angiotensin converting enzyme inhibitors (ACEinh) are known to improve in the short term, cardiac perfusion and function in patients with SSc (Parks JL et al. Rheum Dis Clin North Am 2014). No study has been carried out so far to investigate the long-term effectiveness of these drugs.
Objectives: To address this topic and other aspects of the management of SSc, the DeSScipher (To decipher the optimal treatment of SSc) project was submitted to and financed by the European Community (FP7-HEALTH n°305495).
Methods: From Dec.1, 2012 to Nov. 30, 2015, 512 SSc patients who, as inclusion criterion, had no significant gut or lung or kidney involvement, were treated for 0.5-4 years (median 2.31) with either (n= 359: group 1) vasodilator treatment (i.e. CCB, or ACE-inh or Angiotensin II receptor blockers [AgIIrb] or a combination of them i.e. CCB plus either ACEinh or AgIIrb) or (n= 153:group 2) no vasodilator therapy. The 296 patients of the 2 groups, who had been assessed at baseline and along the follow-up for conduction blocks (CB), ventricular arrhythmias (VA), pacemaker implantation (PMI), left ventricular ejection fraction (LVEF) < 55%) and congestive heart failure (CHF), and sudden cardiac death (SCD) during follow-up, were investigated for the cumulative incidence rate (IR) of all these events. The IR of single manifestations were calculated in the patients investigated for each of them. Cox regression analysis was carried out in the 296 patients with no missing value to identify independent predictors of the occurrence of any myocardial manifestation.
Results: During 1164 patient-years follow-up,6/508 were implanted a PM,10/506 developed a LVEF<55%, 2/492 a CHF,11/325 a VA, 36/305 a CB, none underwent a SCD, any of these manifestations intervening in 59 out of the 296 investigated for all the manifestations. The IR of PMI and VA was greater in group 2 patients as compared with group 1 (1.3 vs 0.2x100 pts/year; p=0.02) and (2.7 vs 1.1x100 pts/year; p=0.077). In stepwise Cox regression analysis, male gender (HR 2.44, 95%CI 1.3-4.6; p=0.005), age at enrollment (HR 2.97, 95%CI 1.7-5.2; p=0.0002) and vasodilator therapy (HR 0.41, 95%CI 0.2-0.7; p=0.002) were independent positive and respectively, negative predictors of new onset cardiac manifestations.
Conclusion: Long term vasodilator therapy is likely to have a preventative role on the occurrence of myocardial manifestations of SSc.
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SAT0255 EVALUATION OF THE CARDIOVASCULAR INVOLVEMENT OF SYSTEMIC SCLEROSIS USING NON-INVASIVE CARDIAC IMAGING TECHNIQUES

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Background: Systemic sclerosis (SSc) is a systemic disease that may affect many organs; among them, cardiac involvement. The prevalence of cardiac involvement in SSc varies depending on the sensitivity of the methods used for its detection. Indirect evidence suggests that subclinical cardiac involvement may eventually occur in the vast majority of patients with SSc. Early detection and monitoring of myocardial involvement are integral to SSc management, as cardiovascular involvement is known to be a poor prognostic indicator when present

Objectives: To describe myocardial perfusion abnormalities and potentially associated coronary arteries lesions using non-invasive imaging techniques in a group of patients with SSc and suggestive symptoms of myocardial involvement (symptomatic) in comparison with a control group of patients with SSc without cardiac symptoms

Methods: A retrospective observational study was performed including a total of 61 patients diagnosed with SSc, 52 symptomatic, with dyspnea and/or chest pain (57.98 ± 12.3 years, 45 women) and 9 asymptomatic controls (50.2±15.21 years, 8 women).

All patients underwent a post-stress (treadmill or pharmacological) myocardic perfusion gated-SPECT, a cold-induced stress SPECT, that were compared to a rest SPECT (to assess ischemia and/or necrosis), as well as a cardiac CT-angiography (to asses significant coronary arteries lesions, considering stenosis of more than 50%)

Results: Twenty-one out of the 52 symptomatic patients (50%) showed myocardial perfusion defects in the stress-rest SPECT: 13 (25%) showed ischemia, 13 (25%) fibrosis/necrosis, and 5 (9.6%) ischemia and necrosis. In the cold-induced SPECT, 17 patients (32.7%) had myocardial perfusion abnormalities: 10 (19.2%) showed ischemia, 13 (25%) fibrosis/necrosis and 6 (11.5%) ischemia and necrosis.

In the other hand, of the 9 asymptomatic patients only 1 (11%) had ischemia and necrosis in the stress-rest SPECT, being only positive for necrosis in the cold-induced SPECT images.

In the cardiac CT-angiography, 7/52 patients (13.4%) showed significant coronary lesions, 4 (57.2%) of them with perfusion defects in the SPECT images, and 3 (42.8%) without significant perfusion alterations

Of the 9 asymptomatic patients, 1 (11%) had significant coronary lesions, being the same patient who presented perfusion defects in myocardial SPECT images

Conclusion: The gated-SPECT is a sensitive tool for detecting myocardial perfusion alterations, normally with no associated significant coronary lesions, suggesting microvascular abnormalities. In this cohort, myocardial perfusion abnormalities were detected in 50% of symptomatic patients, whereas in only 11% of non-symptomatic patients

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SAT0256 EFFICACY AND SAFETY OF ANTIFIBROTIC THERAPY IN DIFFUSE INTERSTITIAL PULMONARY DISEASE ASSOCIATED WITH SYSTEMIC AUTOIMMUNE DISEASES

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Objectives: Assess the efficacy and safety of antifibrotic agents (pirfenidone and nintedanib) in refractory diffuse interstitial lung disease (DILD) associated with systemic autoimmune diseases (SAD).

Methods: Open observational study in patients with active symptomatic DILD-SAD (evidence of clinical and functional impairment) despite treatment with glucocorticoids (GC) and immunosuppressant therapy.

Results: We included 13 patients (8 women) with a mean age (± SD) of 55 ± 12 years (range, 27-71). The baseline SAD were: eighth systemic sclerosis, one rheumatoid arthritis, one ankylosing spondylitis, one microscopic polyangiitis with pulmonary fibrosis and one interstitial lung disease with autoimmune characteristics (IPAF). Mean time from diagnosis to initiation of antifibrotic therapy was 3 ± 8.2 years (IQR 25-75%: 1.6-9.5). The histopathological patterns found were: seven cases (54%) of usual interstitial pneumonia (UIP), three (23%) nonspecific interstitial pneumonia (NSIP), two cases (15%) combined pattern of pulmonary fibrosis plus emphysema (CPFE), and one (8%) non-classifiable pulmonary fibrosis (without histospecific radiological pattern).

In addition to GC, the previous therapy tested for DIDL was mycophenolate mofetil (MMF) in 77% of the patients, cyclophosphamide (8 intravenous boluses) in 46%, and rituximab (RTX) in 54% (the number of RTX cycles administered were 4 ± 3.3; range, 1-9). Despite this, during the year prior to start the antifibrotic drugs, all patients presented a worsening in the values of % pFVC (-8.77%) and% pDLCO (-12.41%).

Five (38%) patients received pirfenidone at doses of 3 tablets/8 hours (2403 mg/day) and eight (62%) nintedanib (5 at a dose of 150 mg/ 12 hours and 3 at a dose of 100 mg/ 12 hours).

Antifibrotic therapy was administered in combination with MMF (1.5-2 g/ day) in 10 patients (77%) and with rituximab in 3 (23%), in addition to GC at doses between 5 and 20 mg/ prednisone per day. The mean

	Pre-treatment (mean ± SD)	Post-treatment (mean ± SD)	Delta (mean)	p
%pFVC	67.9 ± 15.6	74.2 ± 13.9	+ 5.13	NS
%pTLC	66 ± 10.8	77 ± 15.55	+ 4.86	NS
%pDLCO	37.8 ± 7.22	44 ± 7.32	+ 4.42	NS
Walking (meters)	409 ± 52	415 ± 103	+ 7.57	NS

The response in the Pulmonary function testing (PFT) in these 9 patients categorized according to the definitions of the American Thoracic Society was as follows: A)% pFVC: stabilization in 5 cases and improvement in 4; B)% pDLCO: stabilization in 7 cases and improvement in 3.