as myocardial segments. No difference in progression rate was observed stratifying patients according to disease subset or other clinical parameters.

Conclusion: GLS impairment progressed over a 20-month follow-up period in a cohort of right heart catheterization naive cardiac involvement. Further studies are needed to assess the significance of subclinical heart involvement and its progression in patients with SSc.

Disclosure of Interests: None declared

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**SAT0308**

SCREENING TOOLS FOR PULMONARY ARTERIAL HYPERTENSION (PAH) IN SYSTEMIC SCLEROSIS (SSc): A SYSTEMATIC LITERATURE REVIEW (SLR).

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Background: in SSc, PAH has a high morbidity and mortality burden. Therefore, screening and early detection are pivotal to achieve an early diagnosis of PAH.

Objectives: to search for the following of all screening modalities for SSc-PAH in reference to right heart catheterization as diagnostic gold standard.

Methods: papers from 2 previously published SLRs [22 from Glaude et al (1) - from inception to 19/06/2012 - and 22 from Young et al 2018 – from 20/06/2012 to 02/10/2017] were included. The articles’ database was integrated with a systematic search on Pubmed, EMBASE, Web of Science for papers published from 03/10/2017 to 31/12/2018. A total of 199 papers were reviewed and 32 were finally extracted. Bias risk was assessed through QUADAS2 tool.

Results: 167 papers were excluded from data extraction mainly for PAH screening non as main focus or for non-including SSc patients. The 32 papers extracted presented a low bias risk according to QUADAS2. Screening methods reported were:

- Echocardiographic parameters in 31/32 studies, in particular systolic pulmonary arterial pressure (sPAP) in 22 papers; 40 mmHg was the most frequently used cut-off (in 12/22 papers); sPAP was part of a composite algorithm in 9/22 papers. Among others, tricuspid regurgitation velocity (TRV) was used in 6/31 (as part of composite 5/6) and right atrial pressure (RAP) in 3/31 papers.
- Pulmonary function tests parameters in 23/32 papers, with % predicted Lung diffusion for carbon oxygen (DLco) in 21 papers, with a 50% cut-off in 11/21 and as part of composite algorithm in 13/21 studies. Moreover, walked distance at six minutes walking test was a screening parameter in 3/32 papers.
- Serum biomarkers in 12/32 papers, with anti-centromere antibodies (6/12), NT-proBNP (6/12) and uric acid (5/12) being the most frequently reported.
- Clinical parameters in 15/32 papers, with unexplained dyspnoea in 9/15 and telangiectasias in 5/15 papers.
- Composite algorithms were used in 18/32 manuscripts: among them, DETECT (5/18), ESC/EERS 2009 (4/18) or 2015 (3/18) guidelines, ASIG (2/18) e ITINER-air (1/18). In different cohorts, DETECT and ASIG showed higher sensitivity and negative predictive value than ESC/EERS 2009.

Conclusion: in the literature, the screening of SSc-PAH is largely investigated by echocardiographic parameters. In particular, sPAP and TRV, both as single items or part of a composite algorithm, including also serum biomarkers, clinical and functional parameters, are the most frequent parameters evaluated. (supported by an Actelion Pharmaceuticals unrestricted research grant)

References:


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**SAT0309**

CARDIAC MAGNETIC RESONANCE IMAGING ELEVATED NATIVE MYOCARDIAL T1 IS PREDICTIVE FOR THE DEVELOPMENT OF MYOCARDIAL DYSFUNCTION IN SYSTEMIC SCLEROSIS

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Background: All patients included in the study fulfilled the ACR/EULAR classification criteria for SSc. We prospectively included patients who underwent at least two CMR at 1ST, including native T1 and T2 mapping (which give account for myocardial fibrosis and myocardial edema respectively), left and right ventricles morphology and functional assessment, and Late Gadolinium Enhancement (LGE) as a part of routine follow-up between 2015 and 2019.

Objectives: To evaluate the prognostic value of initial abnormal T1 mapping.

Methods: All patients included in the study fulfilled the ACR/EULAR classification criteria for SSc. We prospectively included patients who underwent at least two CMR at 1ST, including native T1 and T2 mapping (which give account for myocardial fibrosis and myocardial edema respectively), left and right ventrices morphology and functional assessment, and Late Gadolinium Enhancement (LGE) as a part of routine follow-up between 2015 and 2019.

Results: Sixty-three patients underwent at list two CMR during the study period. Forty-three patients were women. Mean age was 52.5±15.5 years old. Follow-up duration between the initial and the follow-up CMR was 14.5±11.5 months. Forty-one had diffuse SSC. The mean native T1 was 1066.8±44.6 ms. Twenty-one patients suffered from cardiac clinical manifestations. Nine patients died during the follow-up. Thirty patients (47.6%) had elevated T1 (ET1) with mean T1 1105.4±36.7 ms at the time of initial CMR. Initial ET1 was clearly correlated with: 1/ alteration of Left Ventricle (LV) Ejection fraction (EF) (r=0.5, p<0.0001) during the study period, 2/LV dilatation at initial screening and follow up (r=0.22, p=0.03 and r=0.02, p=0.00) regarding Right ventricle, initial ET1 was correlated with Initial Right Ventricle (RV) dilatation (r=0.3, p=0.02) but neither with RV volume nor RVEF at follow-up. Interestingly, initial ET1 correlated with pericardial effusion (r=0.3, p=0.003) which is known to be a prognostic feature. Seventeen patients (28%) had the LGE but the ET1 at initial screening and follow up was not correlated with LGE.

Six patients had elevated T2 (ET2) which correlated with initial and follow up LV dilatation (r=0.32, p=0.002 and r=0.5, p<0.0001 respectively) but not with LVEF during the period study. Among other parameters, initial increased BNP was correlated with follow up ET1 LVEF and RVEF (r=0.4, p=0.01; r=0.35, p=0.007; r=0.37, p=0.005 respectively). In the same way, initial Pulmonary Arterial Hypertension (PAH) was correlated with follow up ET1 (r=0.3, p=0.02). Initial ET1 did not correlate with age, sex, cardiovascular risk factors, cardiac manifestations or death.

Conclusion: Assessment of diffuse myocardial fibrosis by native T1 is predictive of the occurrence of cardiac dysfunction at the follow-up as initial ET1 was associated with decreased left ventricular function and LV and RV dilatations). These data highlights the potential role of CMR with T1 mapping in initial screening and at the follow-up and provides new insights in the cardiac SSc follow up strategy.

References:

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