

If the presence of CP or CAC score >28 were considered as subclinical atherosclerosis (SAD), a total of 26 patients (60.5%) were diagnosed. We performed multivariate regression analysis, CIMTm, CIMTmax, low High Density Lipid (HDL), high erythrocyte sedimentation rate (ESR) and age were independent factors for the presence of SAD.

Conclusions: SSc patients often have SAD which is misdiagnosed by CV risk charts. Plaque detection by carotid ultrasonography and CT CACscore are useful to detect SAD. The optimal cut-off point of CACscore in this study is 28. SAD detection would be indicated in elderly, patients with low HDL or high ESR. The measure of CIMT could be useful in some cases.

Disclosure of Interest: None declared

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SAT0341 PILOT STUDY: HOME MANAGEMENT OF ILOPROST IN THE MICROPUMP/24 HOURS, IN PATIENTS SUFFERING FROM SCLERODERMA, FOLLOWED FOR 12 MONTHS

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Background: evaluate the effectiveness on Raynaud's phenomenon and on digital ulcers, safety and side effects, during intravenous infusion of iloprost therapy in patients with diffuse and limited scleroderma

Objectives: the effectiveness of the therapy was assessed at 0 and 12 months, on the basis of daily number of episodes of Raynaud's phenomenon, evaluation of digital ulcers changes of the vascular nail bed to capillaroscopy, evaluation of pulmonary hypertension.

Security according to the appearance of adverse events.

Methods: They were enrolled consecutively, for a period of 12 months, 12 patients, 9 women and 2 men, 9 suffering from diffuse scleroderma (positive Scl70) and 3 suffering from limited scleroderma (positive anticentromere), middle age 52,91 years, mean age of disease 7,8 years, 11 non-smoking. They were subjected to infusion of iloprost 1/2 vial diluted in 25 cc of saline solution to 0,6 ml/h, in micropump Infonde for 24 consecutive hours, for 4 days a month for 12 months, 10 patients with peripheral lines and 2 with central venous access.

Results: of 12 patients with the Raynaud's phenomenon, there was a reduction in the daily number in 4 patients, unchanged in 8 patients. Digital ulcers disappeared in 6 patients, they were unchanged in 5 patients, improved although present in 1 patient. In 7 patients capillaroscopic framework remained unchanged, in 5 patients improved capillary density. 10 patients had no pulmonary hypertension, 2 patients had pulmonary hypertension, no change in the two groups at T0 and T 12. None of the patients experienced side effects during the infusion.

Conclusions: the cyclical infusion therapy iloprost prevented the onset of ulcers in patients at T0 not presented or the appearance of new ulcers; it has contributed to the reduction in the daily number of Raynaud's phenomenon and also allowed to reach the same effective dose of a vial of iloprost to dilute in 50 cc of saline solution, allowing the patient to perform at home therapy with greater compliance. In terms of safety none of the patients experienced side effects or adverse effects

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SAT0342 TRANSTHORACIC ECHOCARDIOGRAPHY TO QUANTIFY PULMONARY VASCULAR RESISTANCE IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: One of the major causes of systemic sclerosis (SSc)-related death is pulmonary arterial hypertension, which develops in 12–15% of patients with SSc and accounts for 30–40% of deaths. Consequently, monitoring of pulmonary arterial pressure (PAP) and resistances is essential in patients with SSc. Abbas formula performed by transthoracic echocardiography (TTE) was reported as a good tool to quantify pulmonary vascular resistances (PVR).

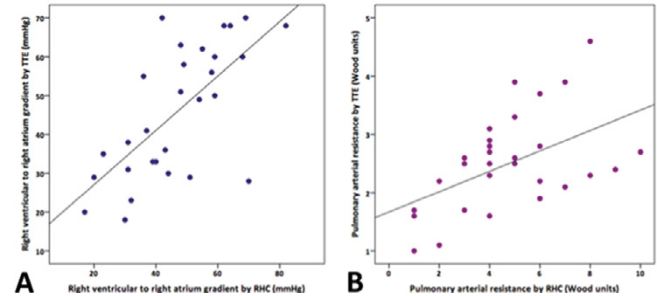
Objectives: Explore the accuracy of TTE and Abbas formula to quantify PVR in patients with SSc.

Methods: All consecutive patients with SSc, diagnosed according to the 2013 ACR/EULAR criteria, or the LeRoy and Medsger criteria for diffuse or limited subsets classification, had within 24h a Doppler echocardiographic examination and right-heart catheterization were performed. The ratio of peak tricuspid regurgitant velocity (TRV, ms) to the right ventricular outflow tract time-velocity integral (TVI_{RVOT}, cm) obtained by Doppler echocardiography (TRV/TVI_{RVOT}) was then correlated with invasive PVR measurements using regression analysis. An equation was modeled to calculate PVR in Wood units (WU) using echocardiography, and the results were compared with invasive PVR measurements [1].

Results: Thirty-three consecutive patients were included, 13 (39.4%) were male and the mean age was 64.6±12.1 years. Most were classified as limited cutaneous SSc (lcSSc; n=29, 87.9%). All patients tested positive for antinuclear antibodies, 18 (21.2%) for anti-scleroderma-70, 7 (54.5%) for anticentromere antibodies and

2 (6.1%) for anti-RNA polymerase III antibodies. Mean and systolic PAP were 31±9 and 53±16 mmHg respectively. There was a good correlation between right ventricle to right atrium gradient pressure assessed by TTE and RHC (R=0.620, P<0.001). RVP assessed by Abbas formula (2.6±1.0 wood units) were well correlated with RVP assessed by RHC (4.8±2.3; R=0.446, P=0.013).

Figure. Correlations between right ventricular to right atrium gradient and pulmonary arterial resistances assessed by transthoracic echocardiography and right heart catheterization



Conclusions: Doppler echocardiography using Abbas formula may provide a reliable, noninvasive method to determine PVR in SSc patients.

References:

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SAT0343 SCREENING OF PULMONARY ARTERIAL HYPERTENSION IN PATIENTS WITH SYSTEMIC SCLEROSIS USING DETECT ALGORITHM – VALIDATION IN THE COHORT OF JAPANESE SINGLE CENTER

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Background: Pulmonary arterial hypertension (PAH) complicated with systemic sclerosis (SSc) has the worst prognosis in PAH associated with other connective tissue diseases (1) and is one of leading cause of death in patients with SSc (2). To improve prognosis in SSc patients, earlier detection and diagnosis of PAH by annual screening is recommended even in asymptomatic patients (3). To effectively detect PAH in patients with SSc at earlier phase, DETECT algorithm is reported as a good tool to identify candidates who need right heart catheterization (RHC), with high sensitivity (4). However, its usefulness has not been validated in Japanese cohorts.

Objectives: To validate the effectiveness of DETECT algorithm in the Japanese single center cohort.

Methods: Patients with SSc who visited Keio University Hospital between 2005 and 2016 were included in the study. Patients over 18 years old, disease duration more than 3 years, and DLCO predicted less than 60% were selected and clinical information was retrospectively collected from records. The sensitivity, specificity, and negative and positive predictive values of the algorithm based on the result of RHC evaluation were calculated in a cohort of PAH patients and non-PAH patients, in whom RHC data were available. Validation with patients with data minimally-required for algorithm were also examined.

Results: Three hundred four cases were visited our hospital during from 2005 to 2016. Patients who fulfilled criteria and had data minimally-required for algorithm were 126 cases. Of 126 patients, 50 were examined RHC evaluation and patients diagnosed as pulmonary hypertension were 26 (21%) and 21 (15%) were PAH. When a cohort of PAH patients and non-PAH patients with RHC data was applied to DETECT algorithm, referral rate to RHC evaluation was 78%, missed diagnosis of PAH was 0%. Sensitivity/specificity for detecting PAH patients were 100%/42%, and positive/negative predictive values were 60%/100%, respectively. Evaluation of 126 patients with data minimally-required for algorithm was also examined. Referral rate to RHC evaluation was 43%, missed diagnosis of PAH was 0%, and sensitivity/specificity and positive/negative predictive values for detecting PAH patients were 100%/72%, 43%/100%, respectively. However, there was a patient who was initially excluded at Step 1, but developed PAH one year later.

Conclusions: The DETECT algorithm was reassured as a good tool to effectively screening tool for PAH in SSc patients. However, we have to keep in mind that unnecessary of RHC referral judged by this algorithm does not guarantee the patient to be free from future development of PAH.

References:

[1] Condliffe R et al. *Am J Respir Crit Care Med* 179:151–157, 2009.

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