Myocardial contrast echocardiography and cardiac magnetic resonance imaging in systemic sclerosis: an observational study of 28 patients

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Introduction
Systemic sclerosis (SS) is a multisystem connective tissue disease characterized by inflammation and excessive collagen deposition leading to diffuse occlusive microvascular lesions and fibrosis. Cardio-pulmonary involvement has become the most important cause of mortality and morbidity.

Patients with SS frequently feature changes secondary to pulmonary hypertension as well as primary myocardial involvement. Primary myocardial involvement is mostly asymptomatic and subclinical. When clinically evident it usually indicates a poor prognosis.

Myocardial contrast echocardiography (MCE) and cardiac magnetic resonance imaging (CMRI) are reliable and sensitive techniques for the assessment of early myocardial involvement.

Objectives
- Evaluate the prevalence of perfusion abnormalities with myocardial contrast echocardiography (MCE) and magnetic resonance imaging (CMRI)
- Identify patterns of perfusion abnormalities and areas of fibrosis
- Investigate the relationship between these changes and left ventricular function

Methods

Population
Inclusion criteria: consecutive patients with SS, followed by the rheumatology department, referred for cardiac evaluation
Exclusion criteria: non-sinus rhythm, systemic arterial hypertension, coronary disease, dipiridamole contraindication

28 consecutive patients (19F, 9M; mean age 53 ± 8 years)

Echocardiogram
- Diastolic dimension of Right Ventricle (RV)
- Left Ventricle (LV) End-Diastolic Volume/mL
- Left Ventricle Ejection Fraction (LV EF)
- LV Regional Function analysis (ASE/AHA model of 16 segments)
- Pulmonary Systolic Arterial Pressure (PSAP)

Results

| Age (mean ± SD) | 52 ± 13 |
| Females (n, %) | 19 (68%) |
| PSAP (mmHg) | 38 ± 6 |
| Diastolic dimension RV (mm) | 20 ± 4 |
| LV EDV (mm³) | 78 ± 11 |
| LV EF (%) | 57 ± 4 |
| Diastolic dysfunction (n, %) | 5 (18%) |
| Wall motion abnormalities (n, %) | 0 (0%) |
| Beta reserve | 1.8 ± 0.5 |
| Diminished Beta-reserve (n, %) | 14 (50%) |
| Qualitative assessment perfusion abnormalities (n, %) | 3 (11%) |
| Late Enhancement (n, %) | 13 (46%) |
| Late Enhancement (grams)* | 4.2 ± 1.8 |

5 patients within normal values (<30mmHg)
23 patients with slight elevation (30-50mmHg)
All patients within normal values (14 - 26mm)
All patients within normal values
All patients within normal values (>50%)
8 patients on the lower limit of normal (50-55%)

Bet reserve was diminished in 17 walls from 14 patients

The correlation we found between the extension of necrosis/fibrosis (LE) and the Beta-reserve suggests that the microcirculation abnormalities are related to necrosis.

No correlation was found between the extension of necrosis/fibrosis (LE) and ejection fraction - possibly due to small sample size.

Conclusions
This study using MCE and CMRI detected perfusion abnormalities and midwall necrosis in the LV that had not been detected by functional studies alone.

These changes might represent a mechanism of LV dysfunction and might influence the prognosis of these patients.

The authors declare no conflict of interest.