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# Circulation

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## Abstract 17650: Myocardial Fibrosis in Systemic Sclerosis, Progression and Response to Therapy Measured by Cardiovascular Magnetic Resonance

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### Abstract

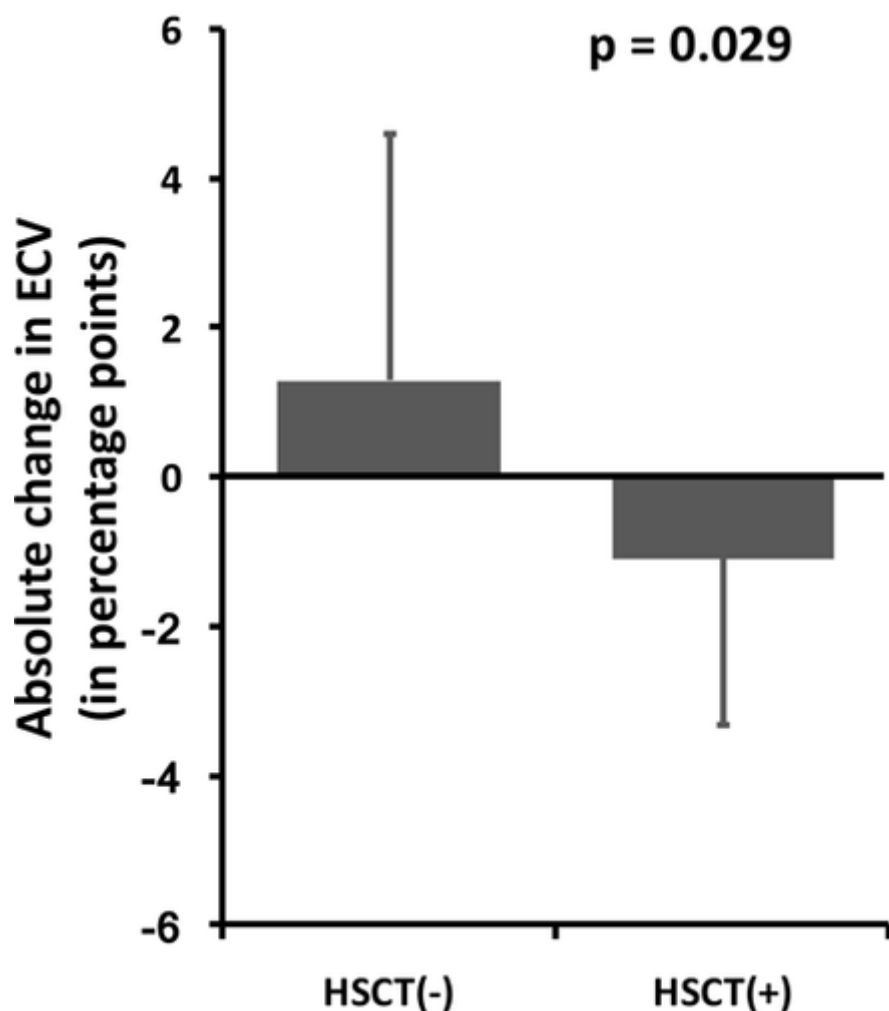
**Introduction:** Myocardial fibrosis is seen in half of Systemic Sclerosis (**SSc**) patients, and is associated with an increased risk of cardiac death. Extracellular volume fraction (**ECV**), an index of myocardial fibrosis measured by cardiovascular magnetic resonance imaging (**CMR**), has been shown to correlate with SSc disease severity measured by the modified Rodnan Skin Score (**mRSS**).

**Hypothesis:** We hypothesized that ECV can quantify progression of myocardial fibrosis and response to therapy in SSc patients.

**Methods:** We identified 30 pairs of CMR exams performed in 21 consecutive SSc patients who were evaluated for hematopoietic stem cell transplantation (**HSCT**). Scan pairs in which HSCT was performed between the first and second CMR were considered HSCT(+), while those without HSCT prior to second CMR were HSCT(-).

**Results:** For all 21 SSc patients (age  $43 \pm 11$ , 86% female), ECV was  $28.9 \pm 2.8\%$  and mRSS was  $16.6 \pm 9.8$  at baseline. Absolute change in ECV was  $+0.1 \pm 3.0\%$  over  $1.2 \pm 0.8$  years in the entire cohort. ECV increased in HSCT(-) pairs ( $n=15$ ) by  $1.3 \pm 3.3$  percentage points over  $0.9 \pm 0.9$  years, while **ECV decreased in HSCT(+) pairs** ( $n=15$ ) by  $1.1 \pm 2.2$  percentage points over  $1.5 \pm 0.8$  years ( $p=0.029$ ). mRSS increased in HSCT(-) pairs by  $4.3 \pm 9.4$  points, but in HSCT(+) patients mRSS decreased by  $10.4 \pm 8.3$  points ( $p < 0.001$ ). Change in ECV correlated with change in mRSS ( $r=0.37$ ,  $p=0.04$ ). In HSCT(+) pairs, native T1 (a sensitive marker of edema) increased by  $30.4 \pm 42.3$  ms ( $p=0.54$ ), suggesting that change in ECV was not due to a reduction in inflammation.

**Conclusions:** Noninvasive quantification of diffuse myocardial fibrosis by CMR measured ECV can detect progression of myocardial fibrosis in SSc patients who have not received HSCT and regression of myocardial fibrosis in SSc patients who have undergone HSCT. The changes in myocardial fibrosis are mirrored by changes in mRSS, a validated measure of disease severity and predictor of prognosis in SSc patients.



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## Footnotes

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