CENTRO HOSPITALAR LISBOA NORTE, EPE

HOSPITAL DE SANTA MARIA

Hospital Pulido Valente

Congresso Novas Fronteiras em Cardiologia

Envolvimento cardíaco na esclerose sistémica - da hipertensão pulmonar à lesão miocárdica

IV Congresso Novas Fronteiras em Cardiologia

Gustavo Lima da Silva

Serviço de Cardiologia I, Hospital de Santa Maria, Centro Hospitalar Lisboa Norte, EPE

- **Sumário**

- **Definição e classificação**
- **Mecanismo de doença**
- **Envolvimento pulmonar**
  - Hipertensão pulmonar
- **Envolvimento cardíaco**
  - Envolvimento cardíaco primário
  - Experiencia do nosso serviço

• Definição e classificação

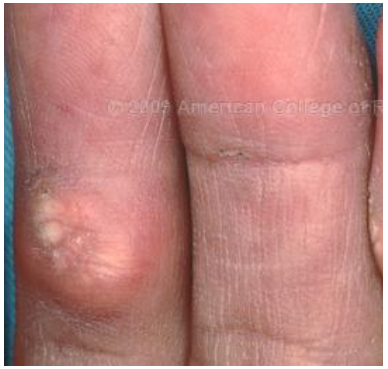
Calcinosis- calcium deposits in the skin

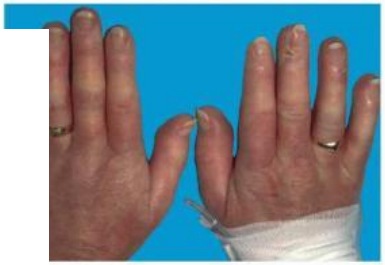
Raynaud's phenomenon- spasm of blood vessels in response to cold or stress

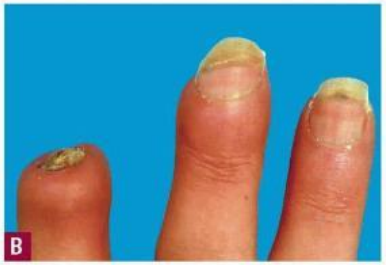
Esophageal dysfunction- acid reflux decrease in motility of esophagus

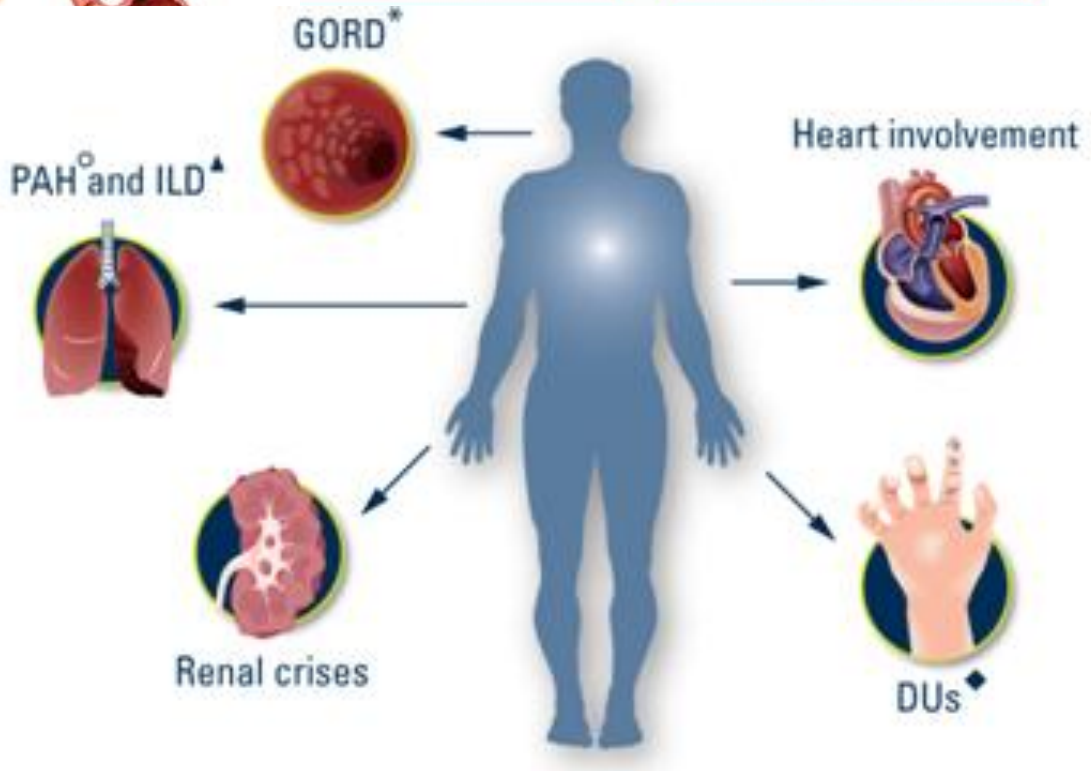
Sclerodactyly- thickening and tight of the skin on the fingers and han

Telangiectasias- dilation of capillaries causing red marks on surface of skin









- **Definição e classificação**

Localizada - Esclerodermia	Sistémica – Esclerose sistémica
1. Linear 2. Morfeia • Localizada • Generalizada	1. Limitada 2. Difusa *3. Sine scleroderma* 4. Exposicional 5. Sobreposição

- **Sumário**

- **Definição e classificação**
- **Mecanismo de doença**
- **Envolvimento pulmonar**
  - Hipertensão pulmonar
- **Envolvimento cardíaco**
  - Envolvimento cardíaco primário
  - Experiencia do nosso serviço

• Mecanismo de doença

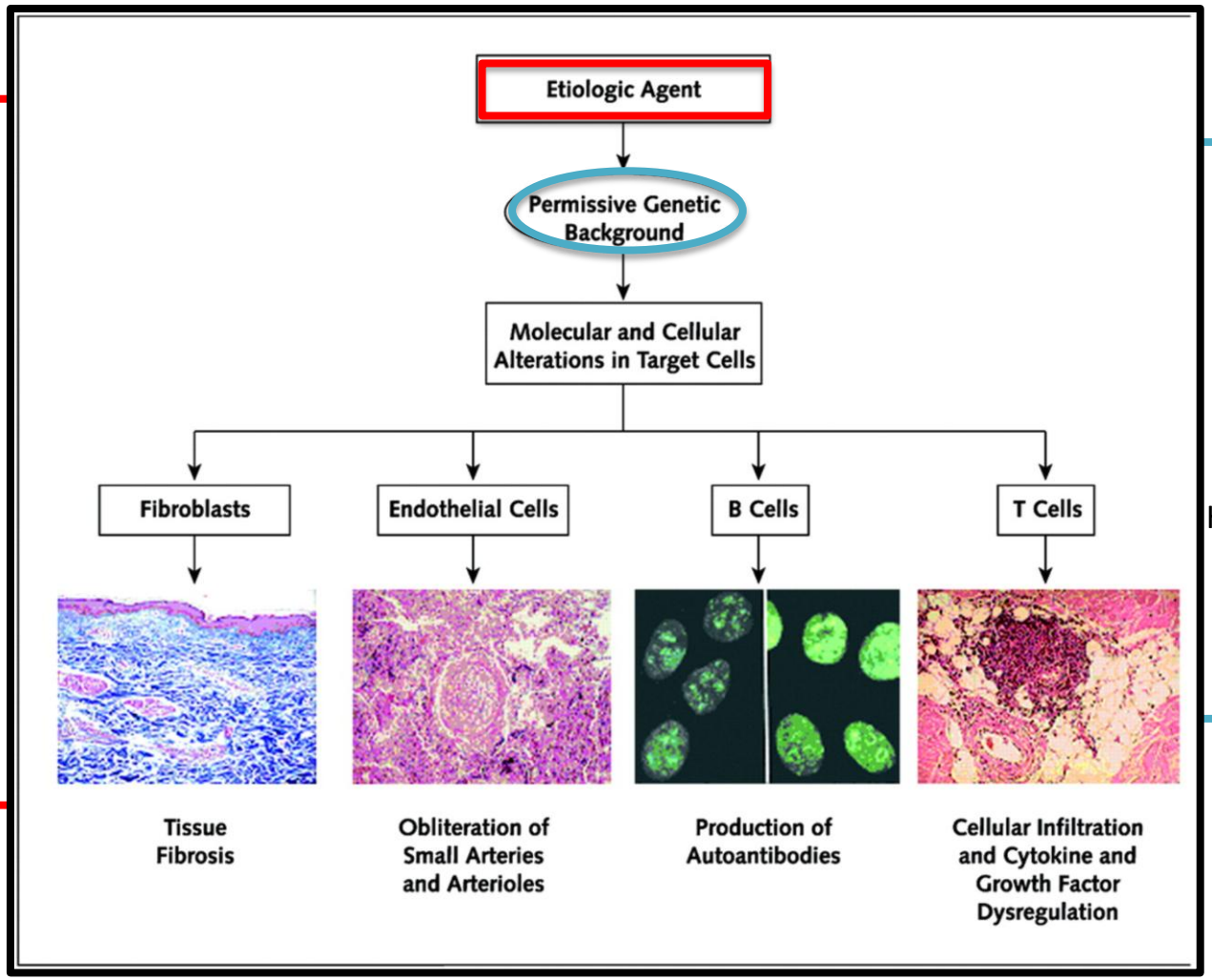
1) Infeccioso
- CMV
- Retrovirus
- Borrelia burgdorferi

2) Não infeccioso

Exposicional
- Derivados petróleo
- Cloreto de vinil
- Oleo de colza
- L-triptofano

Drogas
- Bleomicina
- Docetaxel/paclitaxel
- Cocaina

3) Micro-quimerismo



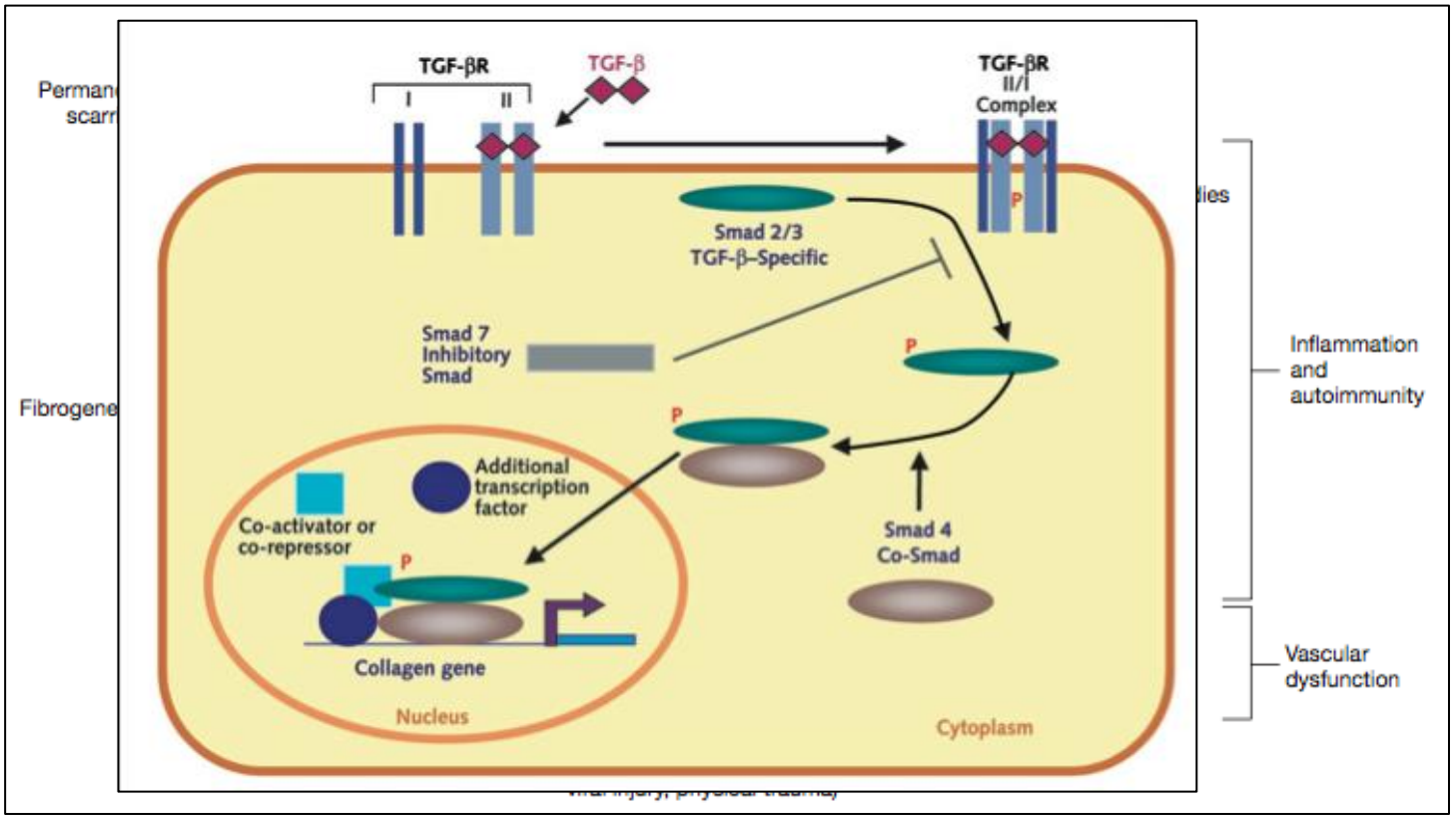
Fibrilina 1
ColA1A
TGF-1

ECA
Endotelina
eNOS

HLA – DQ7, DR2
CD19
IRF5
STAT4
PTPN22

Jimenez, SA., Derk, CT. Following the Molecular Pathways toward an Understanding of the Pathogenesis of Systemic Sclerosis. Ann Intern Med. 2004;140(1):37-50

- **Mecanismo de doença**

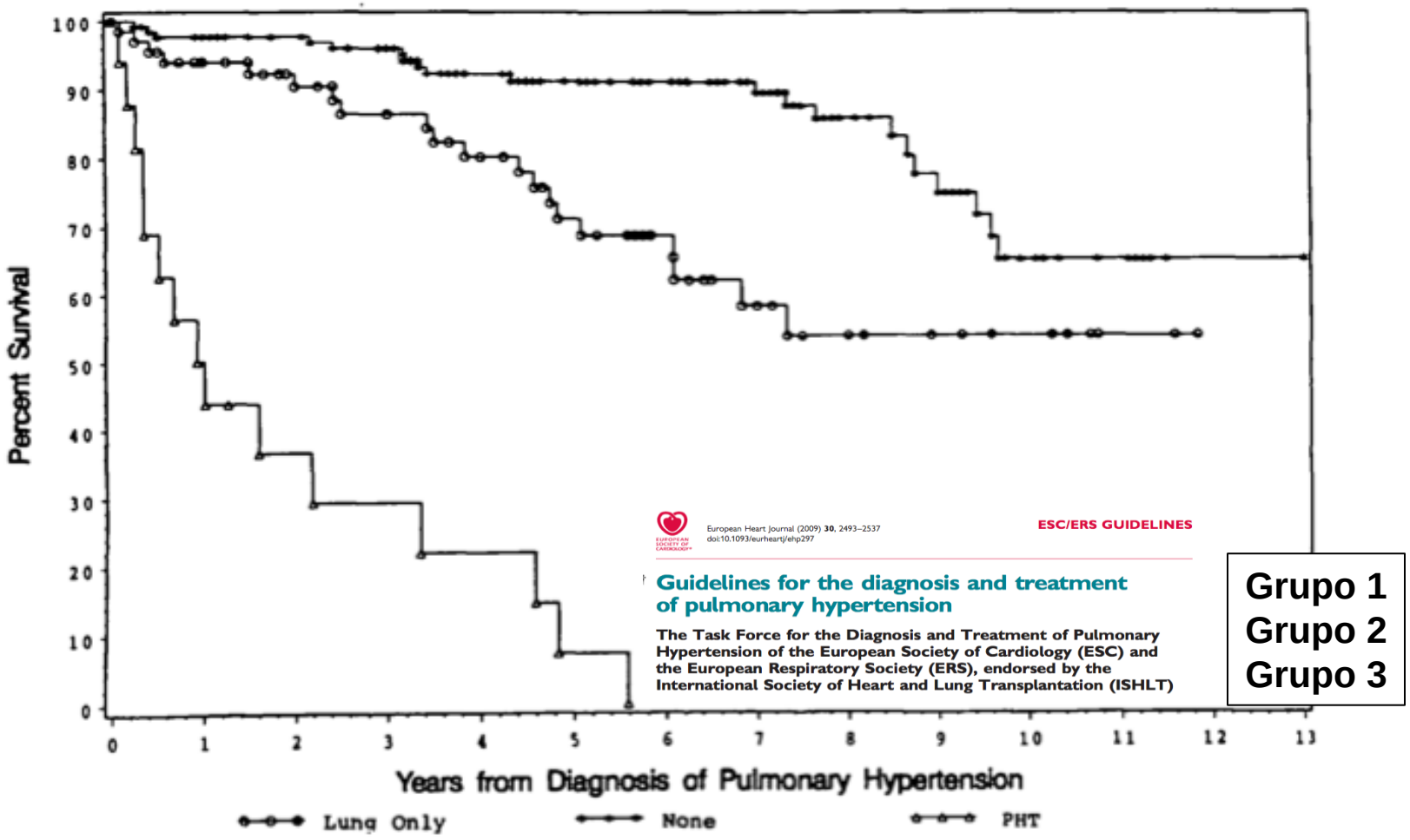


Jimenez, SA., Derk, CT. Following the Molecular Pathways toward an Understanding of the Pathogenesis of Systemic Sclerosis. Ann Intern Med. 2004;140(1):37-50.
Denton, CP., et al: Mechanisms and consequences of fibrosis in systemic sclerosis. Nat Clin Pract Rheumatol. 2006 ;2(3):134-144.

- **Sumário**

- **Definição e classificação**
- **Mecanismo de doença**
- **Envolvimento pulmonar**
  - Hipertensão pulmonar
- **Envolvimento cardíaco**
  - Envolvimento cardíaco primário
  - Experiencia do nosso serviço

- **Envolvimento pulmonar**



Grupo 1
Grupo 2
Grupo 3

Koh, ET., *et al*: Pulmonary hypertension in systemic sclerosis: An analysis of 17 patients. Br. J Rheumatol. 1996; 35: 989 - 998

Arthritis & Rheumatism

Congresso Novas Fronteiras em Cardiologia

Survival and Prognostic factors in systemic sclerosis associated pulmonary hypertension: A systematic review and meta-analysis

Arthritis & Rheumatism 2013: Vol. 65 (9):2412-2423

N = 2.244
Female – 83%
Mean age at inclusion – 60,3 years
lSSc – 79%
dSSc - 18%

Anti-centromere antibodies – 37%
Anti-topoisomerase I antibodies – 13%

NYHA I/II - 23%
NYHA III/IV – 74%

Pericardial effusion - 30%
6 minute walk mean distance – 283m

mPAP - 44mmHg
RAP - 8 mmHg
Cardiac Index - 2,45 L/min/m2
PCWP - 9 mmHg
PVR - 694 dynes/s/cm5
SvO2 - 63%

- The pooled 1-, 2-, and 3-year survival rates were 81% (95% CI 79–84%), 64% (95% CI 59–69%), and 52% (95% CI 47–58%), respectively.
- The 1-, 2-, and 3-year survival rates among patients with PAH were 82% (95% CI 79–85%), 67% (95% CI 63–72%), and 56% (95% CI 51–61%), respectively.
- The 1-, 2-, and 3-year survival rates among patients with ILD-associated PH 77% (95% CI 70–83%), 48% (95% CI 40–55%), and 35% (95% CI 24–47%), respectively

Arthritis & Rheumatism

Survival and Prognostic factors in systemic sclerosis associated pulmonary hypertension: A systematic review and meta-analysis

Arthritis & Rheumatism 2013: Vol. 65 (9):2412-2423

Table 2. Prognostic factors for survival in patients with PAH, patients with ILD-associated PH, and the whole population*

Prognostic factor	PAH	ILD-associated PH	Whole population
Age, years	1.02 (1.01–1.03)	1.02 (0.99–1.04)	1.02 (1.01–1.03)
Male sex	1.57 (1.03–2.40)	0.89 (0.35–2.24)	1.35 (0.95–1.92)
SSc subtype, limited vs. diffuse	0.91 (0.37–2.22)	0.71 (0.41–1.22)	0.70 (0.47–1.03)
Presence vs. absence of ACAs	0.99 (0.23–4.30)	1.12 (0.41–3.03)	1.25 (0.67–2.33)
NYHA functional class III/IV vs. class I/II	2.53 (1.81–3.52)	2.06 (0.82–5.18)	2.47 (1.81–3.37)
6-minute walk distance, per 100-meter increase	0.65 (0.56–0.75)	0.83 (0.63–1.10)	0.68 (0.60–0.78)
Forced vital capacity, % predicted	1.00 (0.99–1.01)	1.02 (1.00–1.04)	1.00 (0.99–1.01)
DLCO, % predicted	0.97 (0.95–0.99)	0.97 (0.94–0.99)	0.97 (0.96–0.98)
Right atrial pressure, mm Hg	1.06 (1.03–1.08)	0.95 (0.89–1.01)	1.02 (0.98–1.05)
Mean PAP, per 10–mm Hg increase	1.27 (1.14–1.40)	1.11 (0.88–1.40)	1.22 (1.11–1.33)
PCWP, mm Hg	0.96 (0.90–1.02)	0.95 (0.85–1.06)	0.96 (0.91–1.00)
Cardiac index, liters/minute/m^2	0.56 (0.53–0.68)	0.95 (0.68–1.33)	0.65 (0.53–0.78)
PVR, dynes · seconds · cm^{-5}	1.07 (1.04–1.11)	0.99 (0.93–1.06)	1.06 (1/02–1.09)
Svo_2, % predicted	0.95 (0.93–0.96)	–	0.95 (0.93–0.97)
Pericardial effusion	1.74 (1.07–2.82)	2.44 (1.03–5.78)	1.88 (1.24–2.87)

* Values are the pooled hazard ratio (95% confidence interval). PAH = pulmonary arterial hypertension; ILD = interstitial lung disease; PH = pulmonary hypertension; SSc = systemic sclerosis; ACAs = anticentromere antibodies; NYHA = New York Heart Association; DLCO = diffusing capacity for carbon monoxide; PAP = pulmonary artery pressure; PCWP = pulmonary capillary wedge pressure; PVR = pulmonary vascular resistance.

- **Sumário**

- **Definição e classificação**
- **Mecanismo de doença**
- **Envolvimento pulmonar**
  - Hipertensão pulmonar
- **Envolvimento cardíaco**
  - Envolvimento cardíaco primário
  - Experiencia do nosso serviço

• Envolvimento cardíaco

Heart involvement is found in more than 90% of patients with SSc and when clinically evident is associated with a 5-year mortality rate of 75%;

Janosik DL, Osborn TG, Moore TL, et al. Heart disease in systemic sclerosis. Semin Arthritis Rheum 1989; 19:191

Primário:

Doença microvascular

Disfunção ventricular

Fibrose miocárdica

Bradidisritmias / taquidisritmias

Regurgitação valvular

Miocardite

Pericardite / derrame pericárdico

Secundário:

Ecocardiografia avançada

Hipertensão sistêmica

Contraste miocárdico

Hipertensão pulmonar

Doppler tecidular

Speckle tracking

Ressonância magnética cardíaca

- **Redução da reserva coronária, disfunção da microcirculação e fibrose miocárdica**

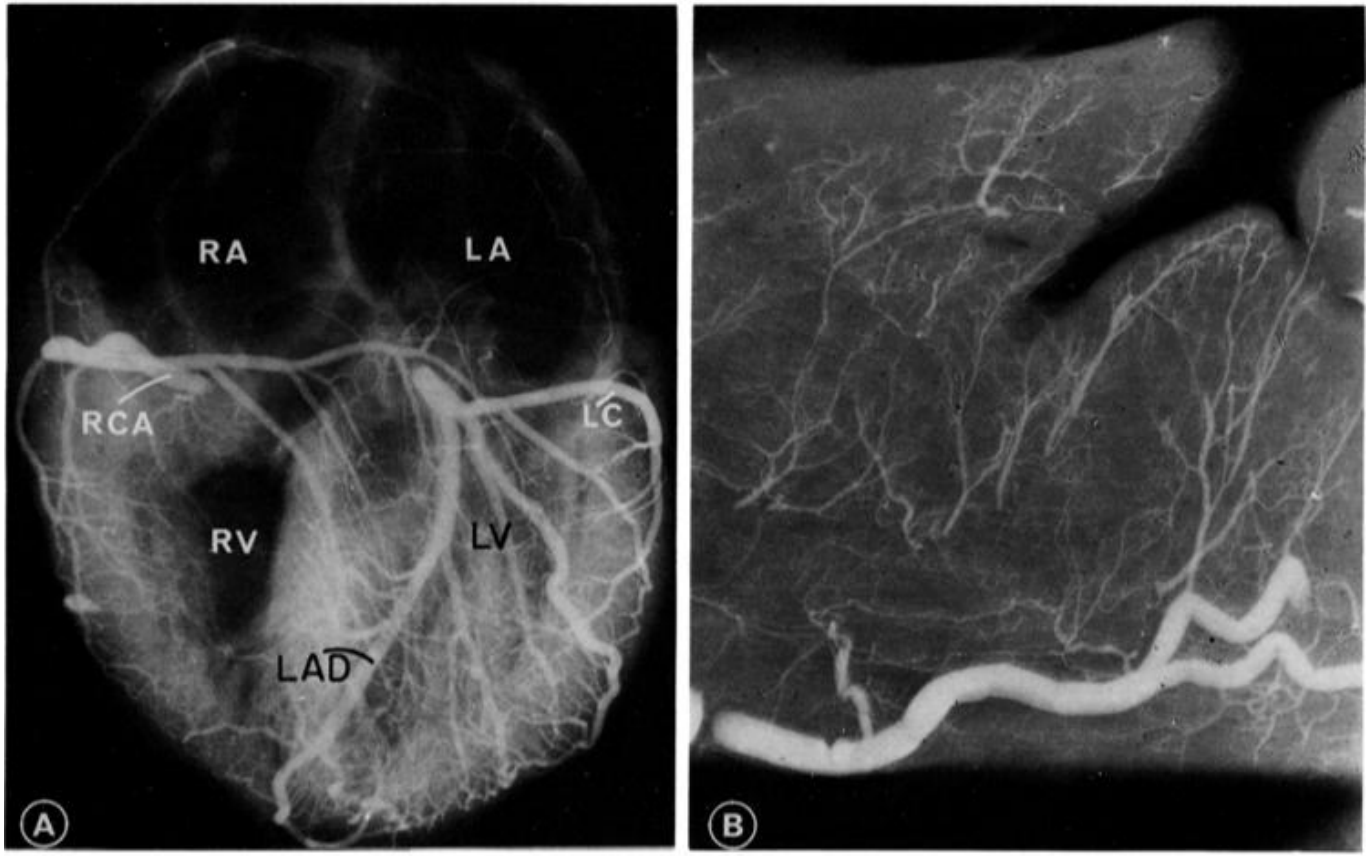


FIGURE 4. *Postmortem arteriogram of heart of patient with PSS showing: A) normal epicardial coronary arteries, and B) normal intramyocardial arterioles on microangiography. RA = right atrium; RV = right ventricle; LA = left atrium, LV = left ventricle; LAD = left anterior descending, LC = left circumflex and RC = right coronary arteries.*

Bulkley, BH., et al.: Myocardial lesions of progressive systemic sclerosis: a cause of cardiac dysfunction. *Circulation* 1976; 53: 483- 490.

- **Redução da reserva coronária, disfunção da microcirculação e fibrose miocárdica**

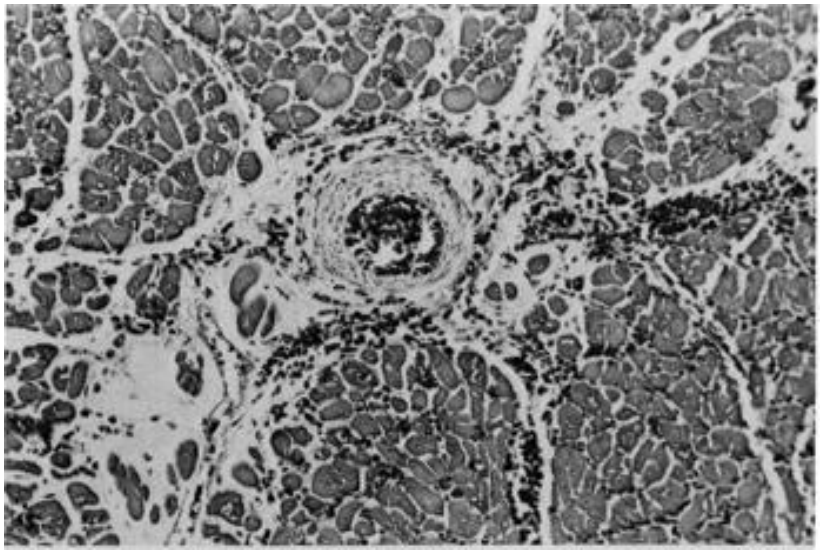
Table 4. Coronary circulation and myocardial metabolism, at baseline and after dipyridamole infusion, in control subjects versus progressive systemic sclerosis (PSS) patients*

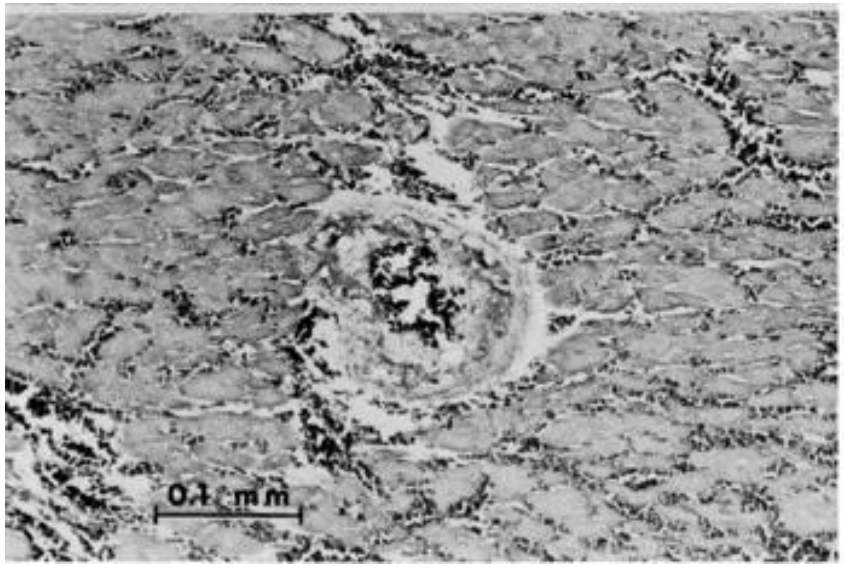
	Baseline			After dipyridamole		
	Control subjects (n = 7)	PSS patients (n = 7)	P†	Control subjects (n = 7)	PSS patients (n = 7)	P
CSBF	100 ± 15	89 ± 32	NS	399 ± 58	191 ± 45	<0.01
CR	0.92 ± 0.10	1.19 ± 0.40	NS	0.21 ± 0.03	0.44 ± 0.11	<0.01
a O_2	17.48 ± 0.95	15.04 ± 1.75	0.02	16.54 ± 0.83	14.26 ± 1.66	<0.02
cs O_2	4.69 ± 0.34	4.50 ± 0.35	NS	12.71 ± 0.37	10.19 ± 1.98	<0.05
a–cs O_2 D	12.56 ± 0.84	10.54 ± 1.46	<0.02	3.51 ± 0.39	4.12 ± 0.64	NS
MVO_2	12.88 ± 2.10	9.06 ± 2.42	<0.01	13.87 ± 1.38	7.86 ± 2.10	<0.01
Lactate extraction	0.62 ± 0.14	0.47 ± 0.22	NS	0.56 ± 0.22	0.54 ± 0.37	NS

* Values are mean ± SD. CSBF = coronary sinus blood flow, ml/minute/100 gm; CR = coronary resistance, mm Hg/ml/minute/100 gm; a O_2 = arterial O_2 content, ml/100 ml; cs O_2 = coronary sinus oxygen content, ml/100 ml; a–cs O_2 D = arterial–coronary sinus oxygen content difference, ml/100 ml; MVO_2 = myocardial oxygen consumption, ml/minute/100 gm.
† NS = not significant.

Kahan, A., *et al*.: Decreased coronary reserve in primary scleroderma myocardial disease. *Arthritis Rheum* 1985; 28: 637-46

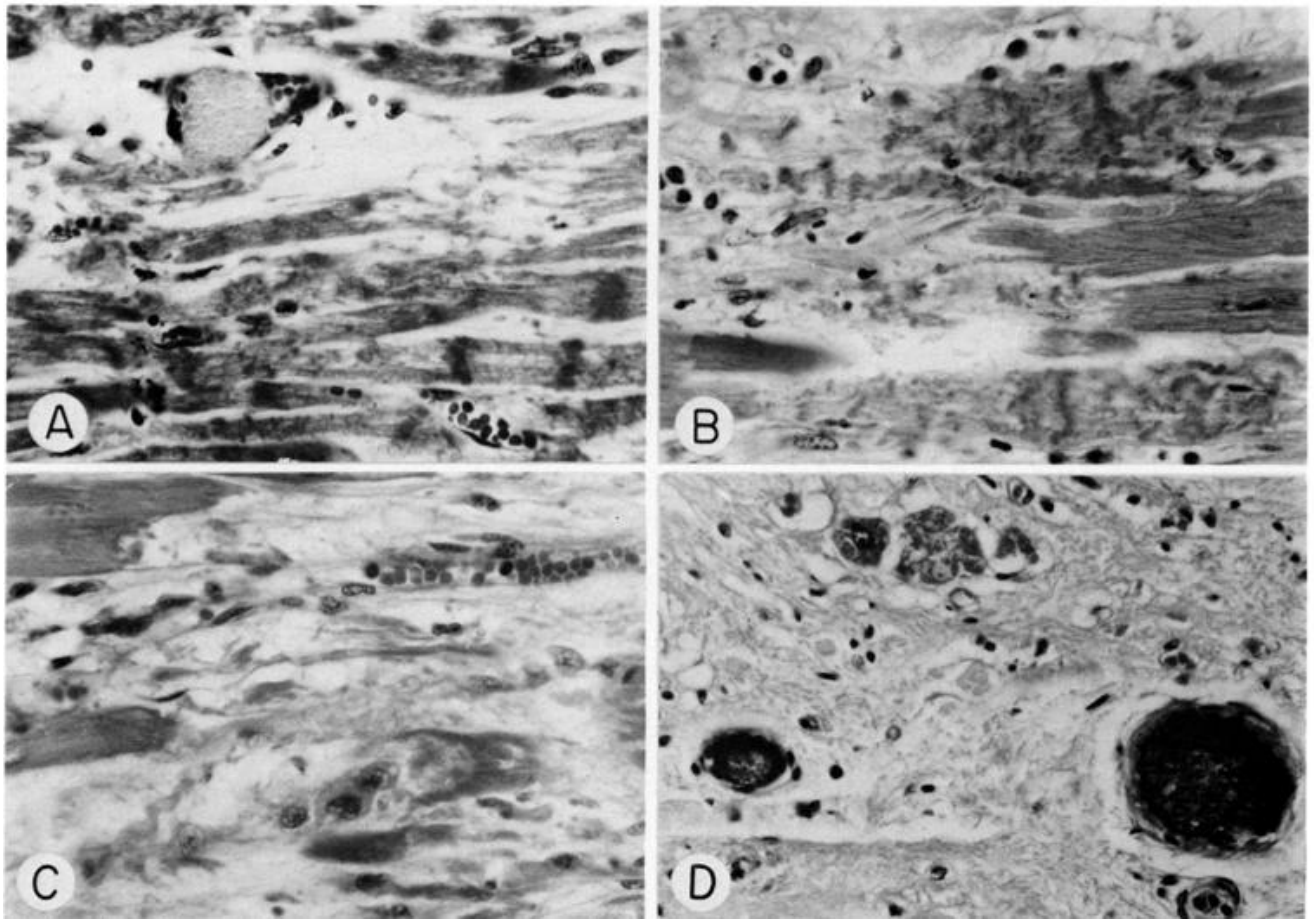
- **Redução da reserva coronária, disfunção da microcirculação e fibrose miocárdica**





James, TN.: De Subitaneis morbitus. VIII. Coronary arteries and conduction system in scleroderma heart disease. *Circulation* 1974; 50: 844-856.

- **Redução da reserva coronária, disfunção da microcirculação e fibrose miocárdica**



Bulkley, BH., et al.: Myocardial lesions of progressive systemic sclerosis: a cause of cardiac dysfunction. *Circulation* 1976; 53: 483- 490.

• **Redução da reserva coronária e disfunção da microcirculação**

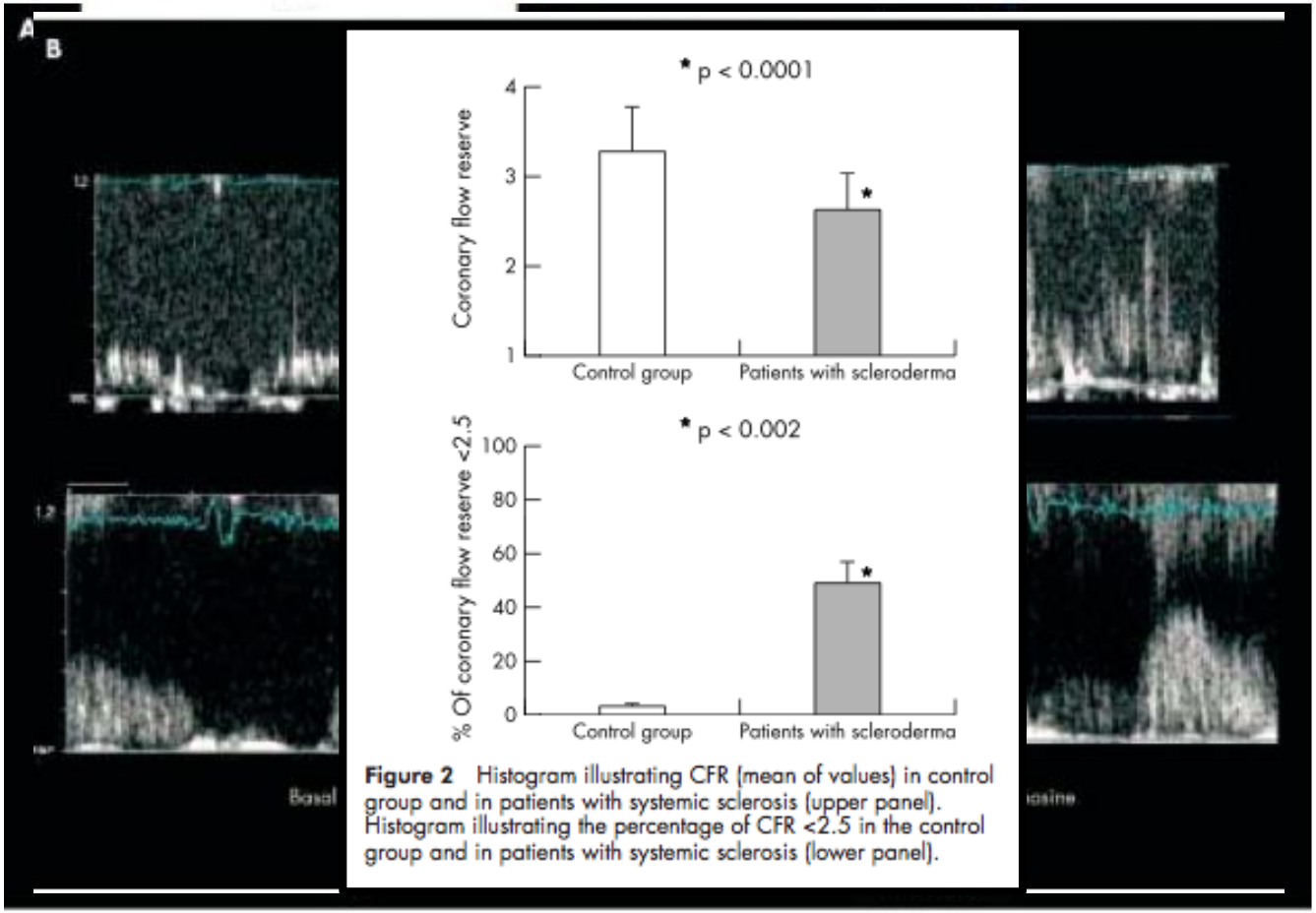


Figure 2 Histogram illustrating CFR (mean of values) in control group and in patients with systemic sclerosis (upper panel). Histogram illustrating the percentage of CFR <2.5 in the control group and in patients with systemic sclerosis (lower panel).

Montisci, R., *et al.*: Detection of early impairment of coronary flow reserve in patients with systemic sclerosis. *Ann Rheum Dis* 2003; 62: 890-3.

• **Redução da reserva coronária e disfunção da microcirculação**

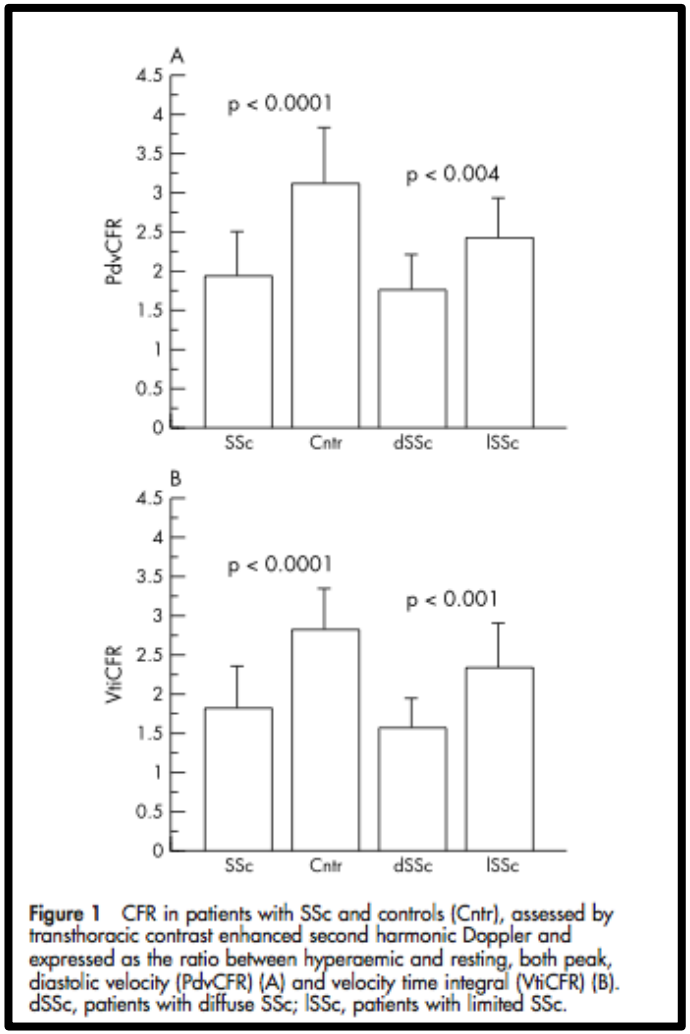
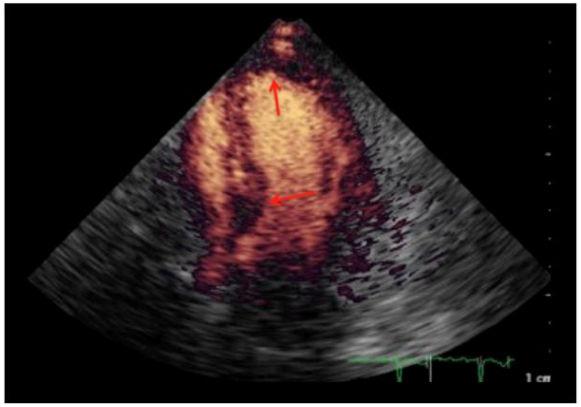


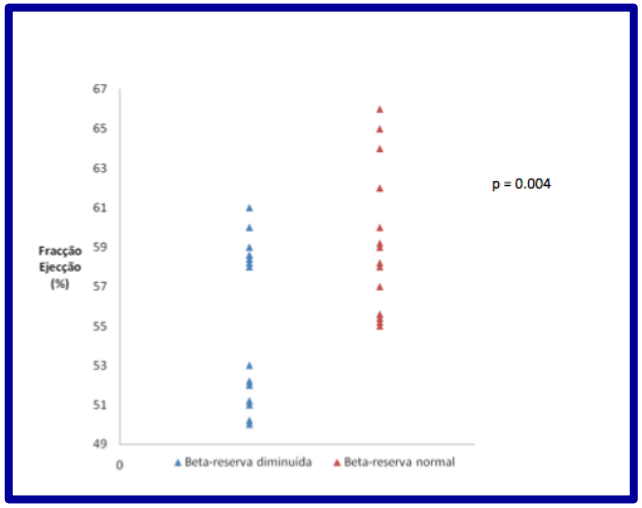
Figure 1 CFR in patients with SSc and controls (Cntr), assessed by transthoracic contrast enhanced second harmonic Doppler and expressed as the ratio between hyperaemic and resting, both peak, diastolic velocity (PdvCFR) (A) and velocity time integral (VtiCFR) (B). dSSc, patients with diffuse SSc; lSSc, patients with limited SSc.

Sulli, A., *et al.*: Blunted coronary flow reserve in systemic sclerosis. *Rheumatology* (Oxford) 2004; 43: 505-9.

Redução da reserva coronária e disfunção da microcirculação na esclerose sistémica
Impacto na função ventricular

Estudar a prevalência e padrão de disfunção da microcirculação e reserva coronária em doentes com esclerose sistémica e determinar a sua relação com a função ventricular esquerda.





(55.1± 4% vs 59.5± 3.8%, p=0.004)

Na nossa população de doentes com ES, a redução da reserva coronária está associada a redução da freacção de ejecção.

• **Disfunção miocárdica**

Follansbee, W.,*et al.*: Physiologic abnormalities of cardiac function in progressive systemic sclerosis with diffuse scleroderma. *N Engl J Med* 1984; 310: 142-8.

Hegedus, I., *et al*: Left ventricular wall motion abnormalities in 80 patients with systemic sclerosis. *Clin Rheumatol* 1995; 14: 161-4.

Valentini, G., *et al.*: Diastolic abnormalities in systemic sclerosis: evidence for associated defective cardiac functional reserve. *Ann Rheum Dis* 1996; 55: 455-60.

Meune, C., *et al.*: High prevalence of right ventricular systolic dysfunction in early systemic sclerosis. *J Rheumatol* 2004; 31: 1941-5.

• **Disfunção miocárdica**

Eur J Echocardiography (2005) 6, 351–357

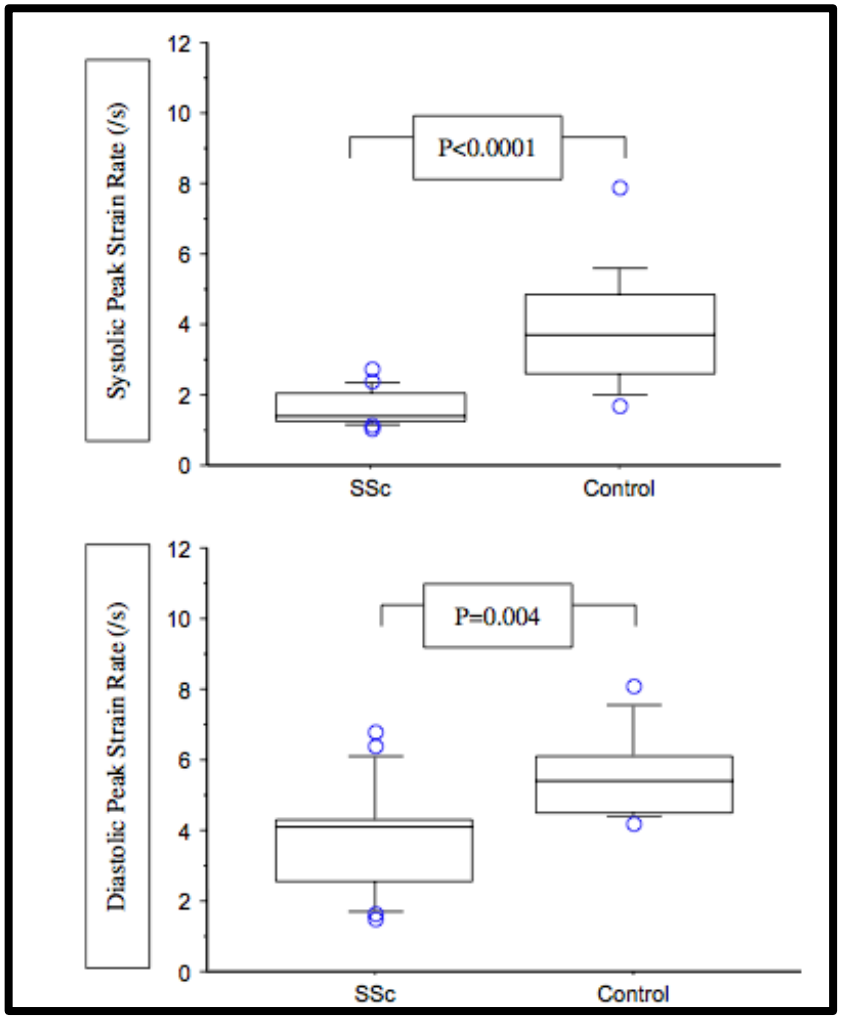
ELSEVIER

EUROPEAN SOCIETY OF CARDIOLOGY®

Myocardial contractility is early affected in systemic sclerosis: A Tissue Doppler echocardiography study

Christophe Meune [a,*], Yannick Allanore [b], Olivier Pascal [c], Jean-Yves Devaux [d], Odile Dessault [d], Denis Duboc [a], Simon Weber [a], André Kahan [b]

- **Disfunção miocárdica**



Meune, C., *et al.*: Myocardial contractility is early affected in systemic sclerosis: A tissue doppler echocardiography study. Eur J Echocardiography 2005; 6: 351-357

2006

Alterações Precoces da Função Ventricular Esquerda na Esclerodermia Detectadas por Doppler Pulsado Miocárdico

M. José Amaro, Ana G. Almeida, Cláudio David, Guilherme Pereira, Catarina Resende, Sónia Ribeiro, Mário G. Lopes

Serviço de Cardiologia – Hospital de Santa Maria
Clínica Universitária de Cardiologia – Faculdade de Medicina de Lisboa

- **Disfunção miocárdica**

European Heart Journal – Cardiovascular Imaging (2012) **13**, 863–870
doi:10.1093/ehjci/jes047

Two-dimensional speckle tracking of the left ventricle in patients with systemic sclerosis for an early detection of myocardial involvement

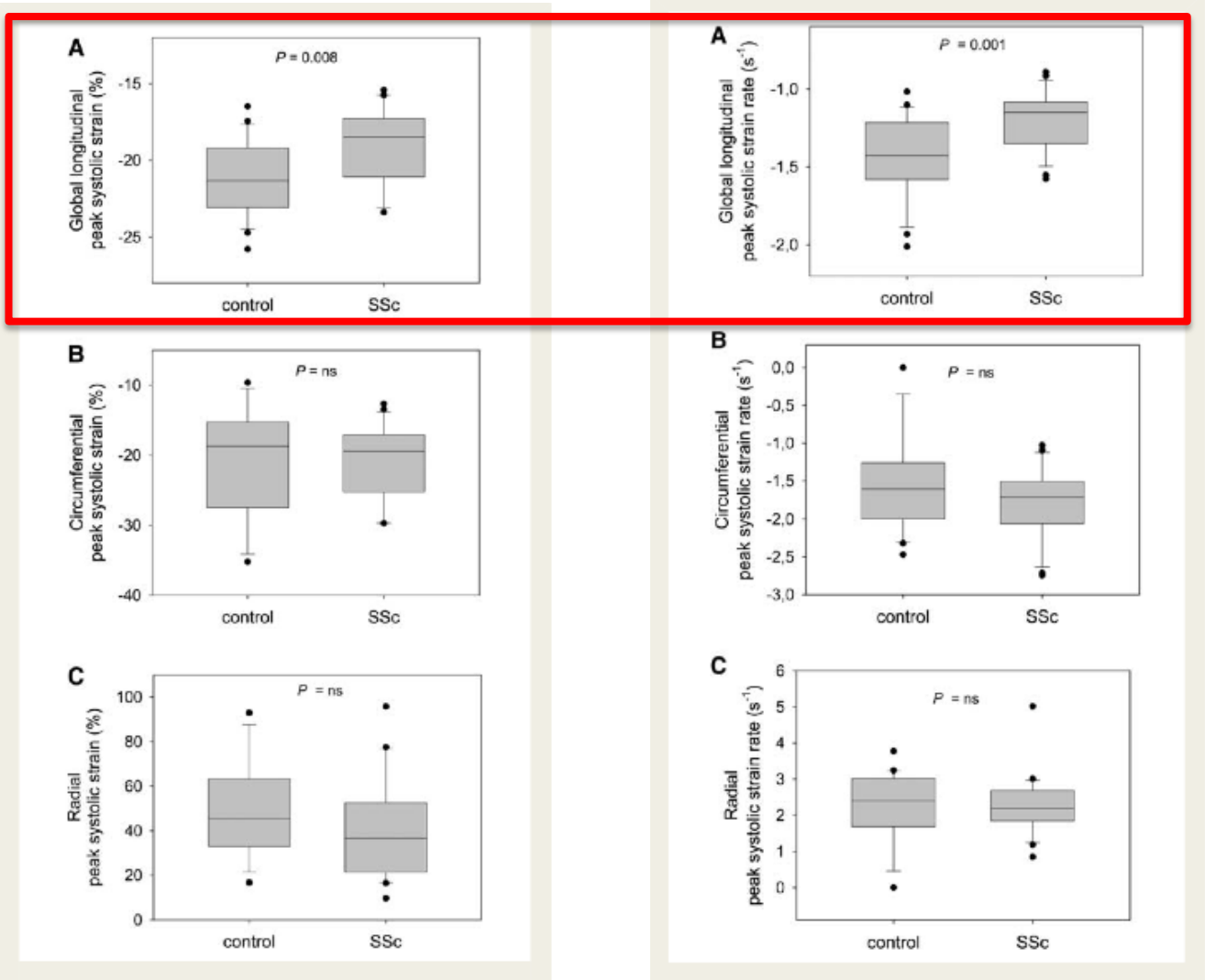
Sebastian Spethmann[1]*, Henryk Dreger[1], Sebastian Schattke[1], Gabriela Riemekasten[2,3], Adrian C. Borges[4], Gert Baumann[1], and Fabian Knebel[1]

[1]Medizinische Klinik für Kardiologie und Angiologie, Campus Mitte, Charité—Universitätsmedizin Berlin, Charitéplatz 1, Berlin D-10117, Germany; [2]Medizinische Klinik mit Schwerpunkt Rheumatologie und Klinische Immunologie, Campus Mitte, Charité—Universitätsmedizin Berlin, Charitéplatz 1, Berlin 10117, Germany; [3]German Rheumatism Research Centre, a Leibniz Institute, Berlin, Germany; and [4]Klinik für Innere Medizin I—Kardiologie, Helios Klinikum, Walterhöferstraße 11, Berlin 14165, Germany

Received 5 October 2011; accepted after revision 9 February 2012; online publish-ahead-of-print 16 March 2012

Spethmann, S., *et al*: Two dimensional speckle tracking of the left ventricle in patients with systemic sclerosis for an early detection of myocardial involvement. Eur Heart J. 2012; 13: 863-870

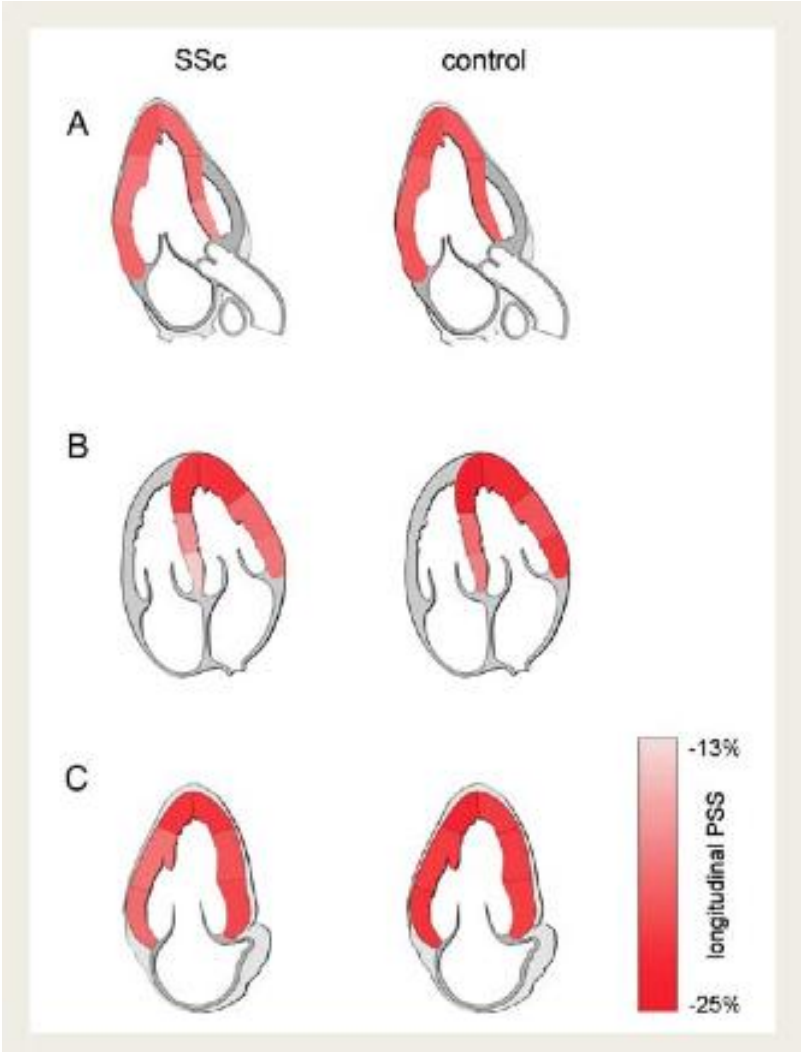
- **Disfunção miocárdica**



Spethmann, S., *et al*: Two dimensional speckle tracking of the left ventricle in patients with systemic sclerosis for an early detection of myocardial involvement. Eur Heart J. 2012; 13: 863-870

- **Disfunção miocárdica**

	SSc ($n = 22$)	Control ($n = 22$)	*P*-value
Longitudinal PSS (%)			
Basal segments	−17.5 ± 3.0	−20.2 ± 2.9	0.004
APLAX	−18.0 ± 3.2	−19.8 ± 4.1	0.21
4CH	−16.2 ± 3.7	−19.9 ± 4.6	0.018
2CH	−19.3 ± 3.9	−22.0 ± 3.2	0.039
Medial segments	−18.4 ± 2.8	−20.1 ± 3.2	0.065
APLAX	−18.5 ± 2.2	−19.9 ± 4.5	0.175
4CH	−17.7 ± 3.4	−19.6 ± 3.6	0.173
2CH	−18.8 ± 4.7	−22.0 ± 3.6	0.023
Apical segments	−20.9 ± 4.6	−23.0 ± 4.2	0.155
APLAX	−19.7 ± 5.3	−22.8 ± 5.2	0.054
4CH	−21.6 ± 5.3	−23.8 ± 5.9	0.446
2CH	−21.7 ± 6.5	−23.0 ± 4.2	0.318
Longitudinal PSSR (s^{-1})			
Basal segments	−1.19 ± 0.21	−1.45 ± 0.29	0.003
APLAX	−1.19 ± 0.27	−1.47 ± 0.39	0.015
4CH	−1.09 ± 0.24	−1.44 ± 0.40	0.004
2CH	−1.32 ± 0.26	−1.55 ± 0.32	0.036
Medial segments	−1.10 ± 0.14	−1.27 ± 0.21	0.004
APLAX	−1.08 ± 0.19	−1.27 ± 0.29	0.022
4CH	−1.04 ± 0.17	−1.24 ± 0.25	0.004
2CH	−1.18 ± 0.24	−1.34 ± 0.26	0.088
Apical segments	− 1.51 ± 0.50	−1.73 ± 0.52	0.132
APLAX	−1.23 ± 0.30	−1.55 ± 0.51	0.034
4CH	−1.31 ± 0.31	−1.57 ± 0.47	0.069
2CH	−1.36 ± 0.38	−1.57 ± 0.48	0.187



Spethmann, S., *et al*: Two dimensional speckle tracking of the left ventricle in patients with systemic sclerosis for an early detection of myocardial involvement. Eur Heart J. 2012; 13: 863-870

Early detection of abnormal LV function using 2D-strain speckle tracking in patients with systemic sclerosis and normal ejection fraction

JS Marques, MD, A G Almeida, MD, PhD, C David, MD, MJ Amaro, MD, MC Amaro, MG. Lopes, MD, PhD

University Hospital Santa Maria, Faculty of Medicine of Lisbon University

- **Associação entre disfunção microcirculatória e miocárdica**

THE LANCET

"A cold-induced vasopastic process in the myocardial circulation might contribute to the development of the patchy myocardial fibrosis seen in patients with systemic sclerosis."

Gustafsson, R., *et al.*: Cold- induced reversible myocardial ischaemia in systemic sclerosis. *Lancet* 1989; 2: 475-479.

- **Associação entre disfunção microcirculatória e miocárdica**

European Journal of Heart Failure (2010) **12**, 268–275
doi:10.1093/eurjhf/hfp198

Cardiac Raynaud's phenomenon induced by cold provocation as a predictor of long-term left ventricular dysfunction and remodelling in systemic sclerosis: 7-year follow-up study

Reiko Mizuno[1], Shinichi Fujimoto[1]*, Yoshihiko Saito[2], and Shinobu Nakamura[1]

[1]Department of General Medicine, Nara Medical University, 840 Shijo, Kashihara, Nara 634-8522, Japan; and [2]First Department of Internal Medicine, Nara Medical University, Nara, Japan

Received 28 September 2009; revised 2 November 2009; accepted 4 November 2009; online publish-ahead-of-print 12 January 2010

- **Associação entre disfunção microcirculatória e miocárdica**

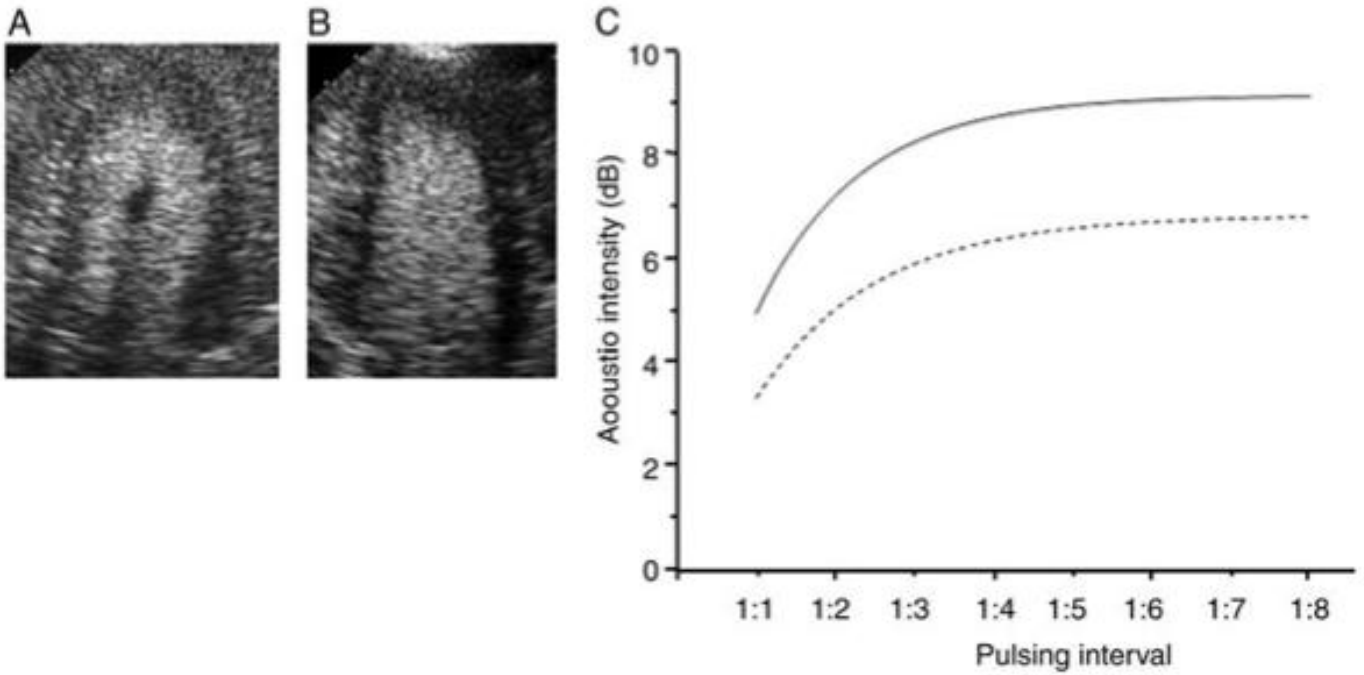


Figure 1 Actual myocardial contrast echocardiography images after cold provocation in a control subject (*A*) and a patient with severe C-Raynaud (*B*) and corresponding plots of acoustic intensity vs. pulsing interval (*C*). Plateau and acceleration rate of intensity were lower in the patient (dashed line) compared with the control subject (solid line). MCE, myocardial contrast echocardiography; C-Raynaud, cardiac Raynauds phenomenon.

Mizumo, R., *et al.*: Cardiac Raynaud's phenomenon induced by cold provocation as a predictor of long-term left ventricular dysfunction and remodelling in systemic sclerosis: 7-year follow-up study. *Eur J Heart Fail* 2010; 12: 268-75.

- **Associação entre disfunção microcirculatória e miocárdica**

Table 3 Comparison of left ventricular ejection fraction and left ventricular end-diastolic volume index at the initial and follow-up evaluation

	Initial evaluation	Follow-up evaluation	*P*-value
LVEF (%)			
All patients ($n = 51$)	67.0 ± 6.2	64.6 ± 8.4	0.005
Patients with C-Raynaud ($n = 15$)	62.9 ± 5.4	55.8 ± 7.2	0.002
Patients without C-Raynaud ($n = 36$)	68.7 ± 5.8	68.3 ± 5.8	0.500
LVEDVI (mL/m^2)			
All patients ($n = 51$)	63.1 ± 13.6	67.1 ± 15.8	<0.001
Patients with C-Raynaud ($n = 15$)	71.5 ± 15.3	82.1 ± 16.7	<0.001
Patients without C-Raynaud ($n = 36$)	59.6 ± 11.3	60.8 ± 10.2	0.178

SSc, systemic sclerosis; ARB, angiotensin receptor blocker; ACE, angiotensin-converting enzyme; C-Raynaud, cardiac Raynaud's phenomenon; LVEF, left ventricular ejection fraction; LVEDVI, left ventricular end-diastolic volume index; PAP, pulmonary artery pressure; MBF, myocardial blood flow; MBV, myocardial blood volume.

Mizumo, R., *et al.*: Cardiac Raynaud's phenomenon induced by cold provocation as a predictor of long-term left ventricular dysfunction and remodelling in systemic sclerosis: 7-year follow-up study. *Eur J Heart Fail* 2010; 12: 268-75.

- **Fibrose miocárdica**

- Cardiac magnetic resonance is the gold standard in identifying myocardial fibrosis.

• **Fibrose miocárdica**

Cardiac magnetic resonance imaging in systemic sclerosis: a cross-sectional observational study of 52 patients

A-L Hachulla,[1] D Launay,[2] V Gaxotte,[1] P de Groote,[3] N Lamblin,[3] P Devos,[4] P-Y Hatron,[2] J-P Beregi,[1] E Hachulla[2]

Ann Rheum Dis 2009;**68**:1878–1884.

- **Fibrose miocárdica**

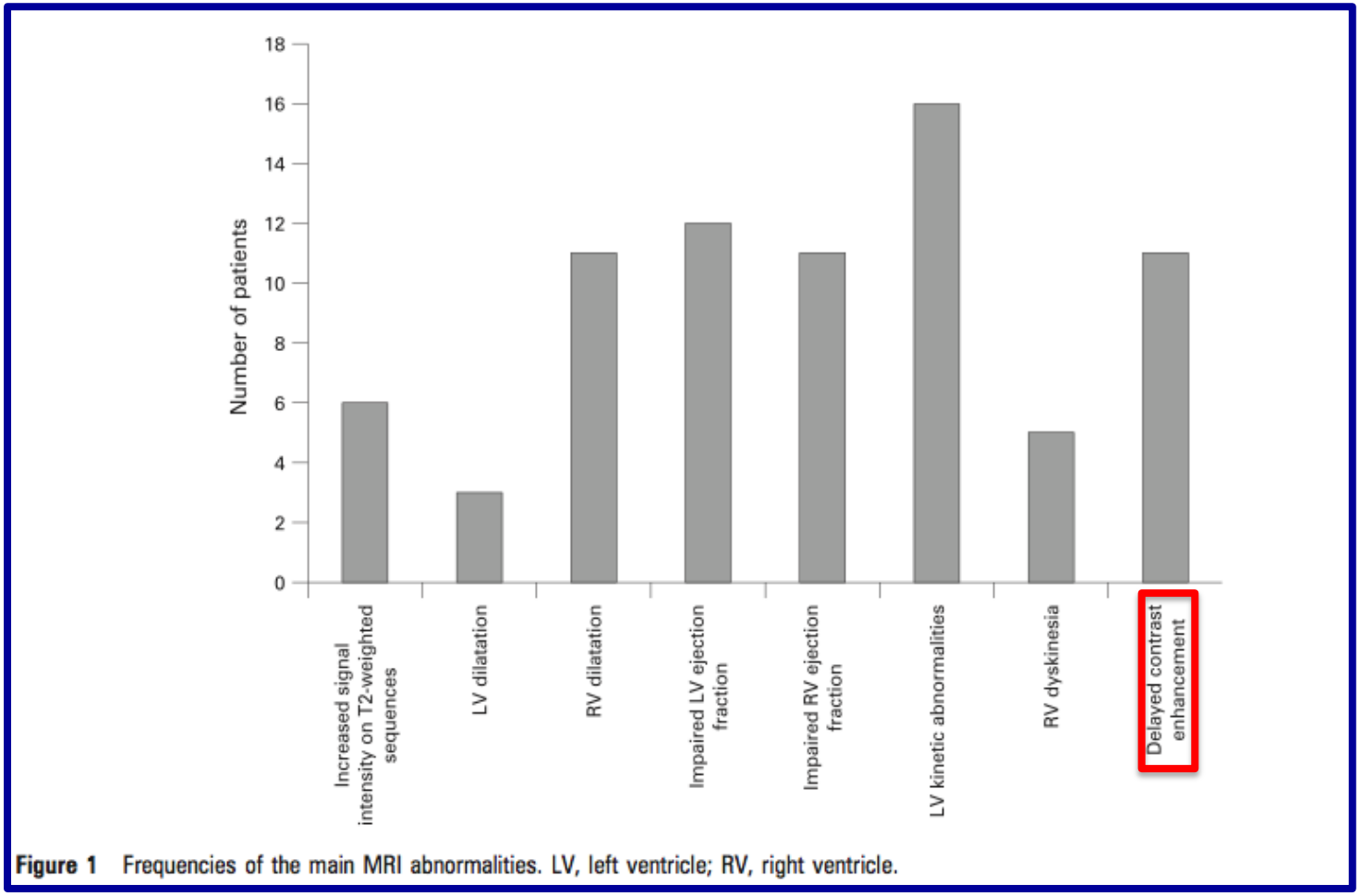


Figure 1 Frequencies of the main MRI abnormalities. LV, left ventricle; RV, right ventricle.

Hachulla, AL., *et al.*: Cardiac magnetic resonance imaging in systemic sclerosis: a cross sectional observational study of 52 patients. *Ann Rheum Dis* 2009; 68: 1878-1884.

- **Fibrose miocárdica**

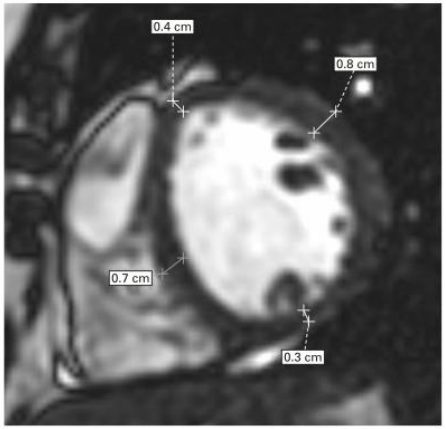


Figure 3 Short axis of the right and left ventricle in cine-MRI sequence showing the thickness of the left ventricle myocardium (end-diastolic frame).

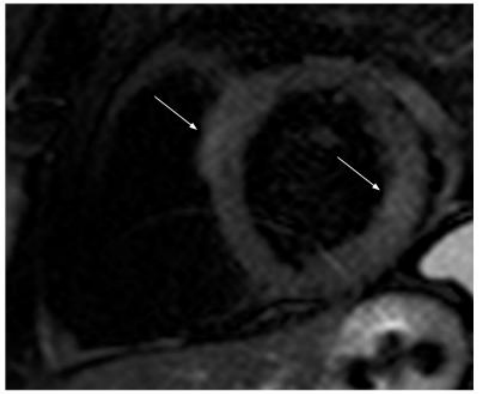


Figure 2 Anteroseptal and lateral (arrows) transmural nodular increased signal intensity in T2-weighted sequence on an MRI 1.5 Tesla scan (Intera, Philips Medical Systems, Best, The Netherlands).

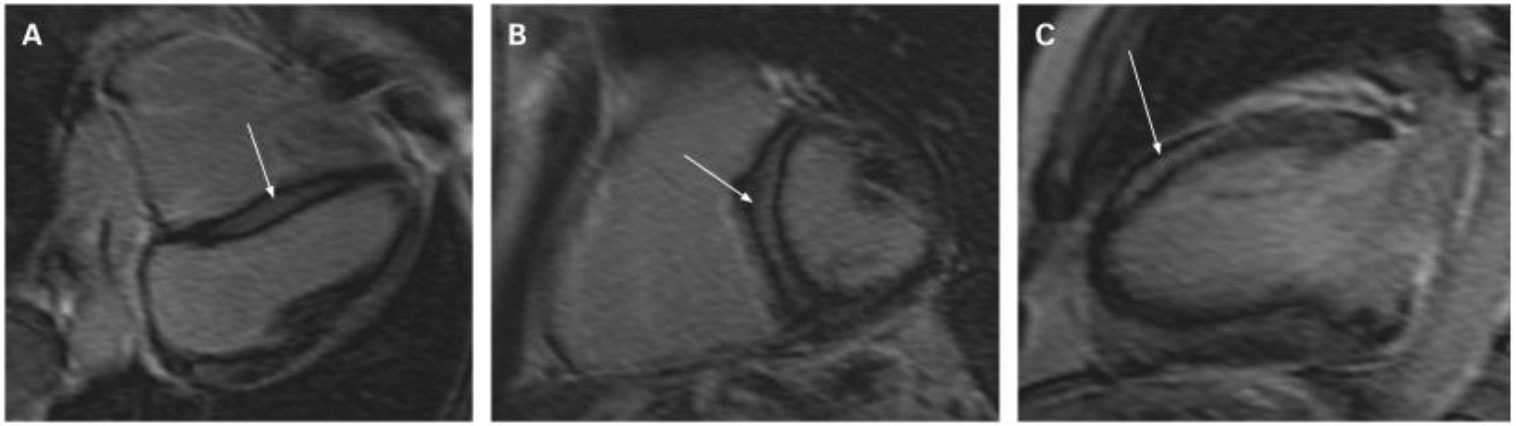


Figure 4 Mid-wall linear late enhancement (arrows) in anteroseptal location assessed by delayed enhancement sequence 10 min after Gd-DTPA injection on an MRI 1.5 Tesla scan. (A) Four chambers; (B) short axis; (C) long axis.

Hachulla, AL., *et al.*: Cardiac magnetic resonance imaging in systemic sclerosis: a cross sectional observational study of 52 patients. *Ann Rheum Dis* 2009; 68: 1878-1884.

- **Fibrose miocárdica**

Table 2 Comparison of cardiac MRI findings between patients with limited cutaneous SSc and patients with diffuse cutaneous SSc

	Limited cutaneous SSc (n = 32)	Diffuse cutaneous SSc (n = 20)	p Value
Patients with at least one cardiac MRI abnormality, n (%)	24 (75)	15 (75)	1.00
Morphological abnormalities			
Increased signal intensity in T2-weighted sequence, n (%)	2 (7)	4 (20)	0.20
Thinned LV myocardium, n (%)	8 (25)	7 (35)	0.44
LV dilatation, n (%)	1 (3)	2 (10)	0.55
RV hypertrophy, n (%)	1 (3)	1 (5)	1.00
RV dilatation, n (%)	5 (16)	6 (30)	0.29
Pericardial effusion, n (%)	7 (22)	3 (15)	0.72
Functional abnormalities			
LV kinetic abnormalities, n (%)	10 (31)	6 (30)	0.92
RV kinetic abnormalities, n (%)	3 (9)	2 (10)	1.00
Mean (SD) LV ejection fraction, %	59 (10)	62 (7)	0.20
Impaired LV ejection fraction	11 (34)	1 (5)	0.02
Mean (SD) LV end-diastolic volumes, ml/m²	68 (14)	69 (17)	1.00
Mean (SD) cardiac output, l/min	4.8 (1.4)	5.6 (1.3)	0.04
Mean (SD) RV ejection fraction, %	52 (12)	50 (10)	0.31
Impaired RV ejection fraction, n (%)	6 (19)	5 (25)	0.73
Mean (SD) RV end-diastolic volumes, ml/m²	73 (11)	87 (28)	0.03
LV diastolic dysfunction on transmitral flow analysis, n (%)	11/29 (38)	4/14 (7)	0.41
Delayed contrast enhancement, n (%)	6 (19)	5 (25)	0.73

Data are mean (SD) or absolute number (%).
LV, left ventricle; RV, right ventricle; SSc, systemic sclerosis.

Hachulla, AL., *et al.*: Cardiac magnetic resonance imaging in systemic sclerosis: a cross sectional observational study of 52 patients. *Ann Rheum Dis* 2009; 68: 1878-1884.

- **Fibrose miocárdica**

Table 4 Comparison of cardiac MRI abnormalities and echocardiography findings

	Normal cardiac MRI (n = 13)	Abnormal cardiac MRI (n = 39)
Normal echocardiography (n = 27)	11	16 (7 thinned LV myocardium, 3 LV kinetic abnormality, 3 impaired RV ejection fraction, 2 impaired LV ejection fraction, 2 LV dilatation, 4 delayed contrast enhancement, 2 increased signal intensity in T2-weighted sequence)
Abnormal echocardiography (n = 25)	2 (abnormal thickness of the interventricular septum or posterior wall of the LV)	23

Hachulla, AL., *et al.*: Cardiac magnetic resonance imaging in systemic sclerosis: a cross sectional observational study of 52 patients. *Ann Rheum Dis* 2009; 68: 1878-1884.

Prevalence of focal myocardial fibrosis in systemic sclerosis assessed by myocardial contrast echocardiography and cardiovascular magnetic resonance. A preliminary study

A G Almeida, MD, PhD, JS Marques, MD, C David, MD, MJ Amaro, MD, MC Amaro, MG. Lopes, MD, PhD

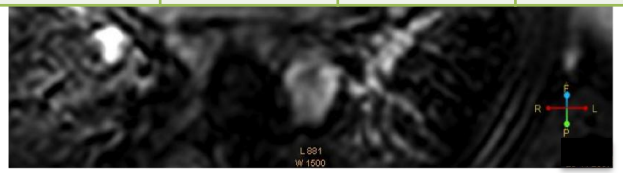
University Hospital Santa Maria, Faculty of Medicine of Lisbon University

MYOCARDIAL FIBROSIS IN SYSTEMIC SCLEROSIS
Impact in ventricular function and clinical presentation

Gustavo Lima da Silva ; Ana G. Almeida; Catarina Resende; João S. Marques; Doroteia Silva; Claudio David; Conceiçao Amaro; Paula Costa, J. A. Pereira Silva, A. Nunes Diogo

To assess the prevalence and patterns of myocardial fibrosis with CMR in patients with SSc and to determine its relationship with clinical presentation and LV function.

	Total	LGE +	LGE -	p value
SSc time course (years; Mean ± SD)	4.3 ± 1.2	4.7 ± 1.2	3.8 ± 1.0	0.040
EF (%; Mean ± SD)	56 ± 8	54.6 ± 6.1	58.9 ± 4.9	0.045



In our population of patients with SSc, CMR detected LV fibrosis, which was associated with the disease duration and LV dysfunction.

Congresso Novas Fronteiras em Cardiologia

- **Relevância do tema**

The Journal of Rheumatology

Meune C., *et al.*: High prevalence of right ventricular systolic dysfunction in early systemic sclerosis. *J Rheumatol* 2004; 31: 1941-1945.

Abstract

OBJECTIVE: To assess right ventricular (RV) function in patients with early systemic sclerosis (SSc) and the acute effects of calcium channel blockers on RV ejection fraction (RVEF).

METHODS: Forty-two consecutive patients with SSc with less than 5 years' disease duration and normal pulmonary arterial pressure (35 women, 7 men; mean age 54.3 +/- 9.7 years; 16 with diffuse and 26 with limited cutaneous forms, systolic pulmonary arterial pressure 30.3 +/- 5.4 mmHg) were prospectively evaluated. All underwent pulmonary function testing, echocardiography, and radionuclide ventriculography at rest and 2 hours after receiving 40 mg oral nicardipine, and were compared at baseline with 20 gender and age matched controls.

RESULTS: None of the patients with SSc had clinical evidence of heart failure. At baseline, SSc patients had significantly lower LVEF (68.5% +/- 7.9 vs 72.4% +/- 5.0, $p = 0.049$) and RVEF (36.5% +/- 7.0 vs 45.8% +/- 5.7, $p < 0.0001$). Sixteen patients had reduced RVEF (< 35%), 3 had reduced LVEF (< 55%), and 10 had reduced peak filling rate (PFR). RVEF correlated to both LVEF and PFR ($r = 0.64$, $p < 0.0001$, and $r = 0.36$, $p = 0.0037$, respectively), whereas no correlation was found with pulmonary function impairment or pulmonary arterial pressure. Nicardipine resulted in a significant increase in RVEF (from 36.5% +/- 7.0 to 42.3% +/- 8.4, $p < 0.001$) whereas afterload indicated by mean arterial pressure did not differ significantly.

CONCLUSION: Reduced RVEF appears to be a common feature in early SSc; it may be due to intrinsic myocardial involvement and is acutely improved by nicardipine.

- **Relevância do tema**

EXTENDED REPORT

Evaluation of the effect of nifedipine upon myocardial perfusion and contractility using cardiac magnetic resonance imaging and tissue Doppler echocardiography in systemic sclerosis

O Vignaux*, Y Allanore*, C Meune, O Pascal, D Duboc, S Weber, P Legmann, A Kahan

Ann Rheum Dis 2005;**64**:1268–1273. doi: 10.1136/ard.2004.031484

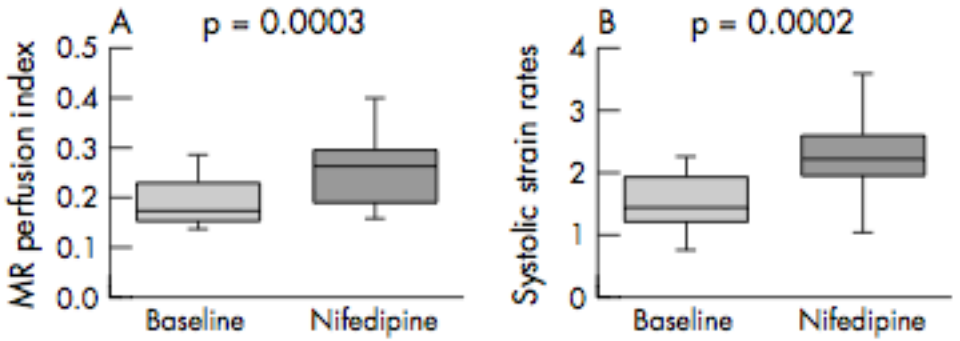
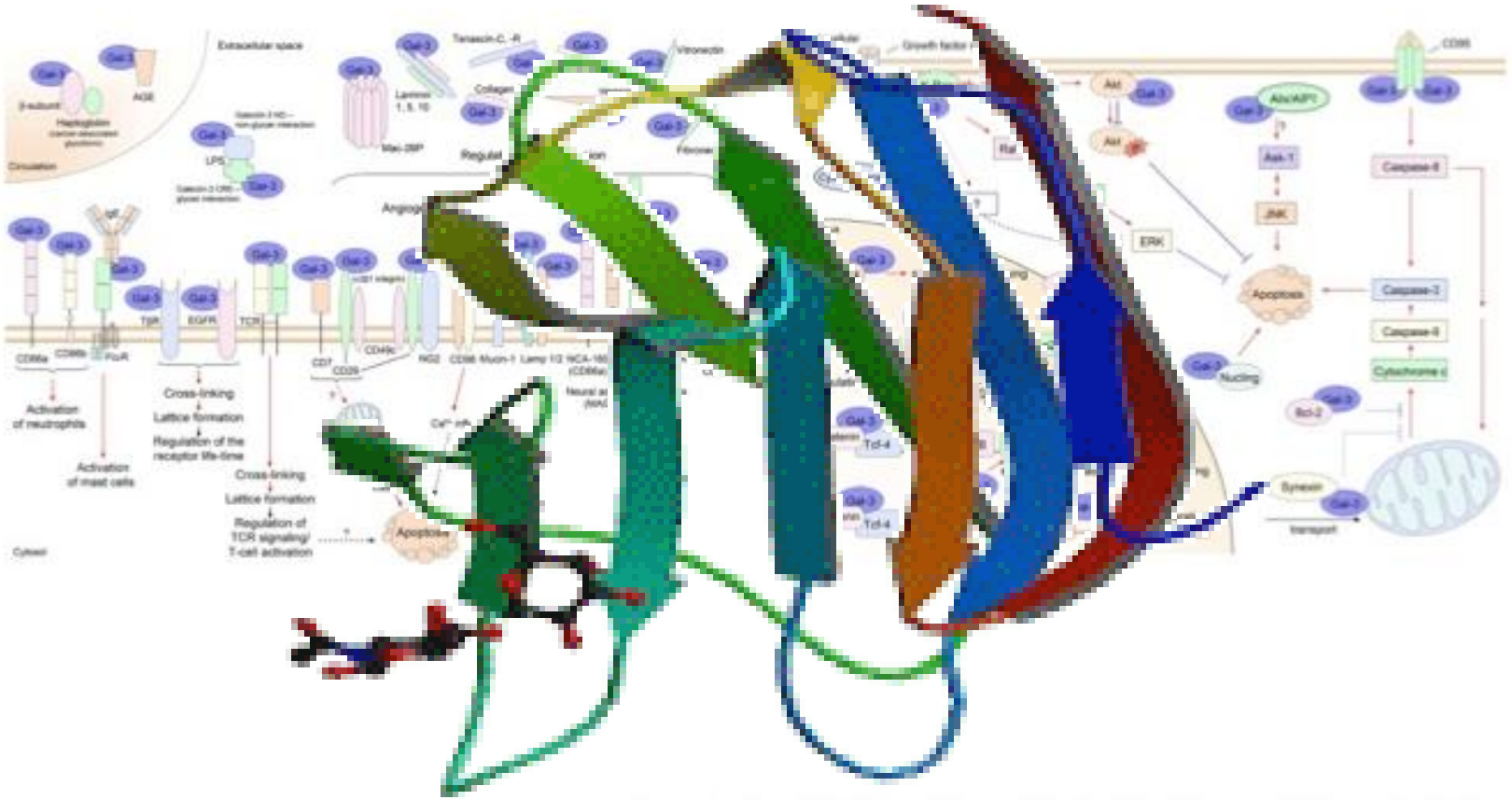


Figure 4 Individual data (25th to 75th centile) of significant increase in myocardial perfusion index, as measured by MRI (A) and increase in systolic strain rate as measured by TDE (B), after 14 days of treatment with nifedipine (60 mg/day).

- **Futuro...**



Dumic, J.; Dabelic, S.; Flögel, M. *Biochim. Biophys. Acta.* **2006**, *1760*, 616.

• **Futuro...**

Envolvimento cardíaco na esclerose sistémica avaliado por imagiologia cardíaca e biomarcadores de fibrose. Estudo de impacto clinico e dos preditores de risco.

Equipa de investigação:

Cardiologia: Gustavo Lima da Silva, João Silva Marques, Susana Martins, Rui Plácido, Paula Costa, Ana G. Almeida

Reumatologia: Cristina Ponte, Alice Castro, Sílvia Fernandes, Catarina Resende

APROVADO PELA COMISSÃO DE ÉTICA DO CHLN EM AGOSTO 2013

INICIO DO RECRUTAMENTO EM JANEIRO DE 2014

CENTRO HOSPITALAR LISBOA NORTE, EPE

HOSPITAL DE SANTA MARIA

Hospital Pulido Valente

Envolvimento cardíaco na esclerose sistémica - da hipertensão pulmonar à lesão miocárdica

IV Congresso Novas Fronteiras em Cardiologia

Gustavo Lima da Silva

Serviço de Cardiologia I, Hospital de Santa Maria, Centro Hospitalar Lisboa Norte, EPE