Supplementary Table S1.

Data extracted for the “Symptoms” domain and its variables, derived from 55 papers.

Symptoms	**N° PAPERS**	**References**	**N° PATIENTS**	**References**
PALPITATION	28	(1-28)	1265 / 4684 (27 %)	from 25 papers (1, 4, 6-10, 14, 15, 17-29)
SYNCOPE	8	(1-4, 6, 16, 19, 30)	9 / 112 (8 %)	from 3 papers (4, 19, 30)
DIZZINESS	5	(2, 3, 6, 26, 31)	10 / 83 (12 %)	from 2 papers (6, 26)
DYSPNOEA	37	(1-3, 5, 7-10, 14-18, 21-24, 26, 28, 30-47)	2178 / 5091 (43 %)	from 28 papers (1, 8-10, 15-18, 21-24, 26, 28, 30-32, 34-36, 40-47)
CHEST PAIN	32	(1-8, 10-13, 15, 17, 20, 21, 23, 24, 26, 30-32, 34, 39, 42, 48-54)	103 / 859 (12 %)	from 23 papers (4, 6-8, 11-13, 15, 17, 20, 21, 23, 24, 26, 30-32, 34, 42, 48, 52-54)
Non specified	5	(37, 55-58)	15/48 (31%)	From 1 paper (55)

References

1. Barison A, Gargani L, De Marchi D, Aquaro GD, Guiducci S, Picano E, et al. Early myocardial and skeletal muscle interstitial remodelling in systemic sclerosis: insights from extracellular volume quantification using cardiovascular magnetic resonance. Eur Heart J Cardiovasc Imaging. 2015;16(1):74-80.
2. Bayar N, Cay HF, Erkal Z, Sezer I, Arslan S, Cagirci G, et al. The importance of fragmented QRS in the early detection of cardiac involvement in patients with systemic sclerosis. Anatol J Cardiol. 2015;15(3):209-12.
3. Bezante GP, Rollando D, Sessarego M, Panico N, Setti M, Filaci G, et al. Cardiac magnetic resonance imaging detects subclinical right ventricular impairment in systemic sclerosis. J Rheumatol. 2007;34(12):2431-7.
4. Di Bello V, Ferri C, Giorgi D, Bianchi M, Bertini A, Martini A, et al. Ultrasonic videodensitometric analysis in scleroderma heart disease. Coron Artery Dis. 1999;10(2):103-10.
5. Fernandes F, Ramires FJ, Arteaga E, Ianni BM, Bonfa ES, Mady C. Cardiac remodeling in patients with systemic sclerosis with no signs or symptoms of heart failure: an endomyocardial biopsy study. J Card Fail. 2003;9(4):311-7.
6. Ferri C, Bernini L, Bongiorni MG, Levorato D, Viegi G, Bravi P, et al. Noninvasive evaluation of cardiac dysrhythmias, and their relationship with multisystemic symptoms, in progressive systemic sclerosis patients. Arthritis Rheum. 1985;28(11):1259-66.
7. Follansbee WP, Curtiss EI, Medsger TA, Jr., Owens GR, Steen VD, Rodnan GP. Myocardial function and perfusion in the CREST syndrome variant of progressive systemic sclerosis. Exercise radionuclide evaluation and comparison with diffuse scleroderma. Am J Med. 1984;77(3):489-96.
8. Follansbee WP, Curtiss EI, Medsger TA, Jr., Steen VD, Uretsky BF, Owens GR, et al. Physiologic abnormalities of cardiac function in progressive systemic sclerosis with diffuse scleroderma. N Engl J Med. 1984;310(3):142-8.
9. Geirsson AJ, Danielsen R, Petursson E. Left ventricular myocardial perfusion and function in systemic sclerosis before and after diltiazem treatment. Scand J Rheumatol. 1996;25(5):317-20.
10. Gurtner C, Werner RJ, Winten G, Krause BJ, Wendt T, Hor G, et al. Early diagnosis of cardiac involvement in systemic sclerosis by 123I-MIBG neurotransmitter scintigraphy. Nucl Med Commun. 1998;19(9):849-57.
11. Kahan A, Devaux JY, Amor B, Menkes CJ, Weber S, Nitenberg A, et al. Nifedipine and thallium-201 myocardial perfusion in progressive systemic sclerosis. N Engl J Med. 1986;314(22):1397-402.
12. Kahan A, Devaux JY, Amor B, Menkes CJ, Weber S, Guerin F, et al. Pharmacodynamic effect of nicardipine on left ventricular function in systemic sclerosis. J Cardiovasc Pharmacol. 1990;15(2):249-53.
13. Kahan A, Devaux JY, Amor B, Menkes CJ, Weber S, Foult JM, et al. Pharmacodynamic effect of dipyridamole on thallium-201 myocardial perfusion in progressive systemic sclerosis with diffuse scleroderma. Ann Rheum Dis. 1986;45(9):718-25.
14. Rodriguez-Reyna TS, Morelos-Guzman M, Hernandez-Reyes P, Montero-Duarte K, Martinez-Reyes C, Reyes-Utrera C, et al. Assessment of myocardial fibrosis and microvascular damage in systemic sclerosis by magnetic resonance imaging and coronary angiotomography. Rheumatology (Oxford). 2015;54(4):647-54.
15. Arakkal G, Chintagunta SR, Chandika V, Damarla SV, Manchala S, Kumar BU. Cardio-pulmonary involvement in systemic sclerosis: A study at a tertiary care center. Indian J Dermatol Venereol Leprol. 2017;83(6):677-82.
16. Meduri A, Di Molfetta DV, Natale L, Manfredi R. Cardiac magnetic resonance in systemic sclerosis patients with cardiac symptoms. Eur Rev Med Pharmacol Sci. 2017;21(21):4797-803.
17. Lee DC, Hinchcliff ME, Sarnari R, Stark MM, Lee J, Koloms K, et al. Diffuse cardiac fibrosis quantification in early systemic sclerosis by magnetic resonance imaging and correlation with skin fibrosis. J Scleroderma Relat Disord. 2018;3(2):159-69.
18. Bosello S, De Luca G, Berardi G, Canestrari G, de Waure C, Gabrielli FA, et al. Cardiac troponin T and NT-proBNP as diagnostic and prognostic biomarkers of primary cardiac involvement and disease severity in systemic sclerosis: A prospective study. Eur J Intern Med. 2019;60:46-53.

19. Thompson AE, Pope JE. A study of the frequency of pericardial and pleural effusions in scleroderma. Br J Rheumatol. 1998;37(12):1320-3.
20. Kitchongcharoenying P, Foocharoen C, Mahakkanukrauh A, Suwannaroj S, Nanagara R. Pericardial fluid profiles of pericardial effusion in systemic sclerosis patients. Asian Pac J Allergy Immunol. 2013;31(4):314-9.
21. Wranicz J, Zielinska M, Cygankiewicz I, Dziankowska-Bartkowiak B, Sysa-Jedrzejowska A. Early cardiovascular involvement in patients with systemic sclerosis (SSc). Med Sci Monit. 2002;8(2):CR78-82.
22. Morelli S, Sgreccia A, Ferrante L, Barbieri C, Bernardo ML, Perrone C, et al. Relationships between electrocardiographic and echocardiographic findings in systemic sclerosis (scleroderma). Int J Cardiol. 1996;57(2):151-60.
23. Follansbee WP, Curtiss EI, Rahko PS, Medsger TA, Jr., Lavine SJ, Owens GR, et al. The electrocardiogram in systemic sclerosis (scleroderma). Study of 102 consecutive cases with functional correlations and review of the literature. Am J Med. 1985;79(2):183-92.
24. De Luca G, Bosello SL, Gabrielli FA, Berardi G, Parisi F, Rucco M, et al. Prognostic Role of Ventricular Ectopic Beats in Systemic Sclerosis: A Prospective Cohort Study Shows ECG Indexes Predicting the Worse Outcome. PLoS One. 2016;11(4):e0153012.
25. Bielous-Wilk A, Poreba M, Staniszewska-Marszalek E, Poreba R, Podgorski M, Kalka D, et al. Electrocardiographic evaluation in patients with systemic scleroderma and without clinically evident heart disease. Ann Noninvasive Electrocardiol. 2009;14(3):251-7.
26. Ferri C, Emdin M, Giuggioli D, Carpeggiani C, Maielli M, Varga A, et al. Autonomic dysfunction in systemic sclerosis: time and frequency domain 24 hour heart rate variability analysis. Br J Rheumatol. 1997;36(6):669-76.
27. Dessein PH, Joffe BI, Metz RM, Millar DL, Lawson M, Stanwix AE. Autonomic dysfunction in systemic sclerosis: sympathetic overactivity and instability. Am J Med. 1992;93(2):143-50.
28. Walker UA, Tyndall A, Czirjak L, Denton C, Farge-Bancel D, Kowal-Bielecka O, et al. Clinical risk assessment of organ manifestations in systemic sclerosis: a report from the EULAR Scleroderma Trials And Research group database. Ann Rheum Dis. 2007;66(6):754-63.
29. Hachulla AL, Launay D, Gaxotte V, de Groote P, Lamblin N, Devos P, et al. Cardiac magnetic resonance imaging in systemic sclerosis: a cross-sectional observational study of 52 patients. Ann Rheum Dis. 2009;68(12):1878-84.
30. Follansbee WP, Zerbe TR, Medsger TA, Jr. Cardiac and skeletal muscle disease in systemic sclerosis (scleroderma): a high risk association. Am Heart J. 1993;125(1):194-203.
31. Oram S, Stokes W. The heart in scleroderma. Br Heart J. 1961;23:243-59.
32. Derk CT, Jimenez SA. Acute myocardial infarction in systemic sclerosis patients: a case series. Clin Rheumatol. 2007;26(6):965-8.
33. Dimitroulas T, Giannakoulas G, Papadopoulou K, Karvounis H, Dimitroula H, Koliakos G, et al. Early detection of cardiac involvement in systemic sclerosis assessed by tissue-Doppler echocardiography: relationship with neurohormonal activation and endothelial dysfunction. J Rheumatol. 2010;37(5):993-9.
34. Candell-Riera J, Armadans-Gil L, Simeon CP, Castell-Conesa J, Fonollosa-Pla V, Garcia-del-Castillo H, et al. Comprehensive noninvasive assessment of cardiac involvement in limited systemic sclerosis. Arthritis Rheum. 1996;39(7):1138-45.
35. Mueller KA, Mueller, II, Eppler D, Zuern CS, Seizer P, Kramer U, et al. Clinical and histopathological features of patients with systemic sclerosis undergoing endomyocardial biopsy. PLoS One. 2015;10(5):e0126707.
36. Gaffney FA, Anderson RJ, Nixon JV, Blomqvist CG. Cardiovascular function in patients with progressive systemic sclerosis (scleroderma). Clin Cardiol. 1982;5(11):569-76.
37. Gottdiener JS, Moutsopoulos HM, Decker JL. Echocardiographic identification of cardiac abnormality in scleroderma and related disorders. Am J Med. 1979;66(3):391-8.
38. Gargani L, Todiere G, Guiducci S, Bruni C, Pingitore A, De Marchi D, et al. Early Detection of Cardiac Involvement in Systemic Sclerosis: The Added Value of Magnetic Resonance Imaging. JACC Cardiovasc Imaging. 2019;12(5):927-8.

39. Mavrogeni S, Karabela G, Koutsogeorgopoulou L, Stavropoulos E, Katsifis G, Plastiras SC, et al. Pseudo-infarction pattern in diffuse systemic sclerosis. Evaluation using cardiovascular magnetic resonance. Int J Cardiol. 2016;214:465-8.
40. Meune C, Allanore Y, Devaux JY, Dessault O, Duboc D, Weber S, et al. High prevalence of right ventricular systolic dysfunction in early systemic sclerosis. J Rheumatol. 2004;31(10):1941-5.
41. De Decker E, Vanthuyne M, Blockmans D, Houssiau F, Lenaerts J, Westhovens R, et al. High prevalence of occupational exposure to solvents or silica in male systemic sclerosis patients: a Belgian cohort analysis. Clin Rheumatol. 2018;37(7):1977-82.
42. Hosoya H, Derk CT. Clinically Symptomatic Pericardial Effusions in Hospitalized Systemic Sclerosis Patients: Demographics and Management. Biomed Res Int. 2018;2018:6812082.
43. Eicher JC, Berthier S, Aho LS, Lorcerie B, Bonnotte B, Laurent G. Measurement of interatrial dyssynchrony using tissue Doppler imaging predicts functional capacity and cardiac involvement in systemic sclerosis. Clin Exp Rheumatol. 2014;32(6 Suppl 86):S-171-6.
44. Karaahmet T, Tigen K, Gurel E, Takir M, Avci A, Cevik C, et al. Impact of systemic sclerosis on electromechanical characteristics of the heart. Heart Vessels. 2010;25(3):223-8.
45. Kostis JB, Seibold JR, Turkevich D, Masi AT, Grau RG, Medsger TA, Jr., et al. Prognostic importance of cardiac arrhythmias in systemic sclerosis. Am J Med. 1988;84(6):1007-15.
46. Dag S, Budulgan M, Dilek B, Batmaz I, Ariturk Z, Nas K, et al. Relation of asymmetric dimethylarginine and cardiac involvement in systemic sclerosis. Acta Reumatol Port. 2014;39(3):228-35.
47. Siegel RJ, O'Connor B, Mena I, Criley JM. Left ventricular function at rest and during Raynaud's phenomenon in patients with scleroderma. Am Heart J. 1984;108(6):1469-76.
48. Bulkley BH, Ridolfi RL, Salyer WR, Hutchins GM. Myocardial lesions of progressive systemic sclerosis. A cause of cardiac dysfunction. Circulation. 1976;53(3):483-90.
49. Yiu KH, Schouffoer AA, Marsan NA, Ninaber MK, Stolk J, Vlieland TV, et al. Left ventricular dysfunction assessed by speckle-tracking strain analysis in patients with systemic sclerosis: relationship to functional capacity and ventricular arrhythmias. Arthritis Rheum. 2011;63(12):3969-78.
50. Alexander EL, Firestein GS, Weiss JL, Heuser RR, Leitl G, Wagner HN, Jr., et al. Reversible cold-induced abnormalities in myocardial perfusion and function in systemic sclerosis. Ann Intern Med. 1986;105(5):661-8.
51. Di Cesare E, Battisti S, Di Sibio A, Cipriani P, Giacomelli R, Liakouli V, et al. Early assessment of sub-clinical cardiac involvement in systemic sclerosis (SSc) using delayed enhancement cardiac magnetic resonance (CE-MRI). Eur J Radiol. 2013;82(6):e268-73.
52. Ellis WW, Baer AN, Robertson RM, Pincus T, Kronenberg MW. Left ventricular dysfunction induced by cold exposure in patients with systemic sclerosis. Am J Med. 1986;80(3):385-92.
53. Gaal J, Hegedus I, Devenyi K, Czirjak L. Myocardial gallium-67 citrate scintigraphy in patients with systemic sclerosis. Ann Rheum Dis. 1995;54(10):856-8.
54. Smith JW, Clements PJ, Levisman J, Furst D, Ross M. Echocardiographic features of progressive systemic sclerosis (PSS). Correlation with hemodynamic and postmortem studies. Am J Med. 1979;66(1):28-33.
55. Steen VD, Follansbee WP, Conte CG, Medsger TA, Jr. Thallium perfusion defects predict subsequent cardiac dysfunction in patients with systemic sclerosis. Arthritis Rheum. 1996;39(4):677-81.
56. Antoniades L, Sfikakis PP, Mavrikakis M. Glucocorticoid effects on myocardial performance in patients with systemic sclerosis. Clin Exp Rheumatol. 2001;19(4):431-7.
57. Giacomelli R, Di Cesare E, Cipriani P, Ruscitti P, Di Sibio A, Liakouli V, et al. Pharmacological stress, rest perfusion and delayed enhancement cardiac magnetic resonance identifies very early cardiac involvement in systemic sclerosis patients of recent onset. Int J Rheum Dis. 2017;20(9):1247-60.
58. Kepez A, Akdogan A, Sade LE, Deniz A, Kalyoncu U, Karadag O, et al. Detection of subclinical cardiac involvement in systemic sclerosis by echocardiographic strain imaging. Echocardiography. 2008;25(2):191-7.