INDIAN HEART JOURNAL 67 (2015) 521–528

Available online at www.sciencedirect.com

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journal homepage: www.elsevier.com/locate/ihj

Original Article

Right ventricular thickness as predictor of global myocardial performance in systemic sclerosis: A Doppler tissue imaging study☆

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ARTICLE INFO

Article history:
Received 9 December 2014
Accepted 12 June 2015

Keywords:
Systemic sclerosis
Doppler tissue imaging
Pulmonary hypertension
Tei index

ABSTRACT

Background: Cardiopulmonary involvement in systemic sclerosis (SSc) is a poor prognostic factor, due to pulmonary hypertension and right ventricular dysfunction. We assessed the echocardiographic parameters of right ventricular (RV) function in SSc and correlated echocardiographic findings to clinical features of the disease.

Methods: Thirty patients with SSc (cases) and 30 healthy, age-matched subjects (controls) were studied. Echocardiography, including tissue Doppler imaging, was used to evaluate cardiac function.

Results: Pulmonary hypertension could be documented in only 5 cases by Doppler echo, using Bernoulli principle. RV diastolic function was significantly deranged in cases. RV systolic function and left ventricle (LV) diastolic function were also significantly deranged in the cases. RV thickness was increased in patients with SSc. There were no significant differences in the echocardiographic variables between diffuse and limited subtypes of SSc. Myocardial performance index (MPI) of both ventricles were increased in cases. We could demonstrate RV thickness as the single most important predictor of MPI of both ventricles with sensitivity of 82% and specificity of 72% for RV-MPI and 63% for LV-MPI. Diastolic function was not found to be affected by disease duration or Rodnan skin score.

☆ Read the Editorial to this manuscript: An Indian-look right into restrictive cardiomyopathies.

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Abbreviations: ACE-I, angiotensin converting enzyme inhibitor; DT, deceleration time; DTI, Doppler tissue imaging; E/A ratio, early diastolic/atrial component velocity ratio; ET, ejection time; FVC, forced vital capacity; Hct, hematocrit; HRCT, high-resolution computed tomography; IVCT/ICT, isovolumic contraction time; ILD, interstitial lung disease; IVRT/IRT, isovolumic relaxation time; LV, left ventricle/ventricular; LVEDD, left ventricular end diastolic dimension; LVEDV, left ventricular end diastolic volume; LVEF, left ventricular ejection fraction; LVESD, left ventricular end systolic dimension; LVESV, left ventricular end systolic volume; MPI, myocardial performance index; PAH, pulmonary arterial hypertension; PAP, pulmonary artery pressure; PASP, pulmonary artery systolic pressure; PAT, pulmonary acceleration time; RR, electrocardiographic R–R interval; RVEF, right ventricular ejection fraction; RV, right ventricle/ventricular; SSc, systemic sclerosis.

http://dx.doi.org/10.1016/j.ihj.2015.06.021

Conclusion: Patients with SSc exhibit abnormal RV and LV diastolic functions as well as abnormal RV systolic function. RV wall thickness was found to be simple and the single best predictor of global myocardial performance. RV dysfunction may be a response to intermittent pulmonary arterial hypertension, lung parenchymal involvement, or secondary to LV diastolic dysfunction in SSc.

1. Introduction

Systemic sclerosis (SSc) is a multisystem disorder characterized by extensive vascular damage and fibrosis of the skin and internal organs. Cardiac involvement occurs in a significant percentage (≤80%) of these patients.[1] Cardiac involvement consists of degeneration of myocardial fibers and irregular areas of interstitial fibrosis that are most prominent around blood vessels.[2]

Myocardial fibrosis seems to start from the subendocardium in SSc. Therefore, due to the subendocardial location of the longitudinal myofibrils, longitudinal functions and diastolic parameters have particular significance in determining early myocardial involvement. The prognosis for patients with SSc also correlates with the presence of pulmonary hypertension.[2]

There is a discrepancy between the frequency of clinically evident myocardial disease (25%) and autoptical myocardial fibrosis (81%).[3] Invasive and non-invasive diagnostic investigations, as well as necropsy studies, showed that, in SSc, cardiac involvement is one of the most frequent visceral complications that can affect the overall prognosis of the disease.[4]

The hemodynamic parameters of right ventricular (RV) dysfunction, as measured by cardiac catheterization, are strong predictors of outcome in patients with SSc.[5,6] Denton et al.[7] have compared Doppler echocardiography vs. right heart catheterization and found Doppler to be 90% sensitive and 75% specific in detecting pulmonary hypertension as compared to catheterization. However, it may underestimate the true PA (pulmonary artery) pressure in absence of tricuspid regurgitation. In the present study, we have applied the technique of tissue Doppler in order to identify the TDI (tissue Doppler imaging) markers of myocardial function.

2. Methods

2.1. Clinical

Seventy-two patients from Rheumatology Outpatient Clinic with diagnosis of SSc according to American College of Rheumatology criteria[8] were screened, and of them, 30 patients were enrolled into the study. Thirty age- and gender-matched healthy volunteers were selected for comparison. Written informed consent was taken from all subjects. The study was conducted for a period of 18 months in Rheumatology clinic of PGIMER. Patients without sinus rhythm, known cardiomyopathies, IHD (ischemic heart disease), significant left-sided valvular disease, CKD (chronic kidney disease) stage III or higher, diabetics, prior cardiac surgery, Bundle branch block on ECG, poor echo window, and those unwilling to give informed consent were excluded.

Detailed history and clinical examination, including Rodnan skin score,[9] were done. Disease duration was calculated based on either onset of symptoms of exertional dyspnea, skin thickening, Raynaud's phenomenon, or physician-aided diagnosis of scleroderma. Pulmonary involvement was defined clinically by either dyspnea or bibasilar rales. Renal function tests, LFT (liver function tests), ANA (antinuclear antobody), Chest X-ray, and pulmonary function tests with diffusion capacity of carbon dioxide (DLCO) were done in all patients.

2.2. Pulmonary function testing

All the patients underwent pulmonary function testing. $FEV_1/FVC < 80\%$ was classified as obstructive. In presence of normal FEV_1/FVC, FVC was used to classify restrictive pattern as mild (<80–60%), moderate (<60–40%), and severe (<40%), as per our laboratory standards. DLCO was adjusted for alveolar ventilation and hematocrit by the following formula.[10] Corrected $DLCO = DLCO \times (10.22 + Hb)/1.7 \times Hb$ .....in males and Corrected $DLCO = DLCO \times (9.38 + Hb)/1.7 \times Hb$ .....in females

2.3. Cardiological investigations

Resting ECG was performed in all. Echocardiography was done using ultrasound [Acuson Sequoia (C512)] equipped with a 4.0-MHz (V4c; Acuson) transducer and DTI technology. The examination was performed with the subject in the left lateral decubitus position with normal breathing. All tracings were recorded at the end-expiratory phase to reduce respiratory variation. Standard two-dimensional parasternal and apical projections were obtained. All recordings were performed with a simultaneous superimposed phonocardiogram to detect the pulmonary component of the second heart sound (S2) and continuous EKG for pulmonary valve flow recording. All images were recorded at sweep speeds of 50 mm/s and 100 mm/s.

From M-mode echocardiography recordings, measurements were made on consecutive three beats, according to recommendations of the American Society of Echocardiography.[11]

The left ventricle (LV) mass was calculated using Devereux formula.

$$\text{Myocardial mass} = 1.04[(\text{LVID} + \text{PWT} + \text{IVST})^3 - \text{LVID}^3] \times 0.8 + 0.6$$

where LVID is the internal dimension of LV, PWT is posterior wall thickness, IVST is interventricular septal thickness, and