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Long term effects of bosentan treatment in patients with early systemic sclerosis

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Introduction: Systemic sclerosis (SSc) is a chronic connective tissue disease characterized by endothelial dysfunction and fibrosis of the skin and internal organs. Also SSc is associated with significant disability and depressive symptoms. The etiology of SSc is currently an expanding area of study. Raynaud's Phenomenon (RP), digital ulcers (DU) and pulmonary artery hypertension (PAH) are the most common manifestations of SSc. Endothelin-1 plays a key role in the pathogenesis of SSc. Bosentan is a dual ET-1 receptor antagonist approved for the treatment of the PAH and to prevent the onset of new DU. The aim of the present study was to investigate the effects of bosentan on exercise capacity, echocardiographic parameters and on quality of life in patients with early-SSc.

Materials and Methods: Ten female patients (61±19 years) with SSc with at least 1 active DU were studied. At study baseline, all patients received bosentan at a dosage of 62.5 mg twice a day orally for 1 month; after liver function control, the dosage was increased to 125 mg twice a day. All patients were followed for 12 months. At baseline and at the twelve month were performed: six-minute walking test (6MWT); Doppler transthoracic-echocardiography; SF-36 self-administered questionnaire for quality of life assessment. Significant level was set at $p < 0.05$.

Results: The 6MWT distance increased, from 385, 89±90, 37 to 437, 86±111, 00 meters; ($p < 0.001$); the right systolic function evaluated by TAPSE increased from 22, 70±3, 62 mm to 25, 38±4, 07; ($p = 0.001$) and the TRV_{max} decreased from 25, 13±9, 21 mm to 21, 66±6, 70; ($p = 0.003$). Finally, statistically significant improvements were found in the scores of SF-36 on following domains: bodily pain (+28%), general health perception (+17%), vitality (+22%) and mental health (+16%).

Conclusions: The study suggests that long-term therapy with bosentan in early-SSc patients can improve exercise capacity, right ventricle performance and quality of life.

Biography

Luca Fallavollita is an investigator of Italian National Institute on Aging. He did his medical school and received his cardiologic training at the University of L'Aquila. He is about to finish his Ph.D. in Cardiovascular Sciences. His scientific interests are focused on Pulmonary Arterial Hypertension and autonomic cardiac regulation. He has a number of presentations in national and international conferences and has published few papers on this regard.

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