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Prognostic value of pulmonary artery elastic properties in patients with pulmonary hypertension - a comparison of Eisenmenger syndrome to other types of pulmonary hypertension

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Patients with Eisenmenger's syndrome (ES) have better survival than other patients with pulmonary arterial hypertension (PAH) probably due to the preservation of right ventricular (RV) function. As in PAH patients RV remodeling and function depend not only on pulmonary artery (PA) pressure but also on the intrinsic properties of PA wall, there is also a possible role of PA stiffness (PAS) as outcome predictor in this setting. Purpose. To study the prognostic role of PAS parameters assessed by 2D transthoracic echocardiography in patients with ES compared to other patients with pulmonary hypertension (PH) receiving specific vasodilator therapy. Methods. Sixty-eight PH patients were enrolled: 27 ES patients and 41 non-ES patients, including patients with other types of PAH (12 idiopathic PAH, 5 operated congenital heart disease, 10 connective tissue disease, 7 other forms of PAH) or chronic thromboembolic PH (7 patients) receiving oral vasodilator therapy. Clinical data, B-type natriuretic peptide (BNP), RV function and PAS parameters were assessed: pulmonary capacitance (PC), PC indexed to body surface area (PC/BSA), pulsatility, elastic modulus (EP), beta-index. PH patients were followed-up for 2.9 years (4 months-6.8 years). Results. Pulmonary vascular resistance (PVR) assessed by right heart catheterization was similar in both groups (11.9 ± 8.0 vs 11.0 ± 6.4 Wood units, p = 0.68). ES patients had lower BNP levels (lnBNP 3.63 ± 1.31 vs 5.31 ± 1.33, p < 0.001) and better RV function than non-ES patients: RV-free wall S wave, RV-S (12.2 ± 2.3 vs 10.2 ± 2.0 cm/s, p < 0.001), RV fractional area change, RV-FAC (40 ± 7 vs 32 ± 9%, p < 0.001), RV global longitudinal strain (RV-GLS) on 3 segments (-20.2 ± 4.4 vs -14.8 ± 6.0%, p = 0.001) or 6 segments (-16.2 ± 4.2 vs -13.1 ± 4.9%, p = 0.011). In ES patients PAS parameters were less impaired than in non-ES group (PC 1.68 ± 0.86 vs 1.18 ± 0.66 ml/mmHg, p = 0.014; PC/BSA 1.05 ± 0.53 vs 0.68 ± 0.37 ml/mmHg m2, p = 0.003; pulsatility 18.8 ± 8.4 vs 13.8 ± 6.4%, p = 0.007, EP 390.7 ± 198.6 vs 578.8 ± 341.6 mmHg, p = 0.007; beta index 6.09 ± 2.85 vs 10.77 ± 6.21, p < 0.001). During follow-up, 12 cardiac deaths occurred: 1 in ES group and 11 in non-ES group (p = 0.021). In non-ES group, predictors of cardiac death were parameters of RV function and PAS: BNP levels (lnBNP 6.20 ± 1.10 in deceased patients vs 4.97 ± 1.27 in survivors, p = 0.007), RV-S (9.1 ± 2.0 vs 10.6 ± 1.9 cm/s, p = 0.038), RV-FAC (25 ± 8 vs 35 ± 7%, p = 0.001), RV-GLS on 3 segments (-11.1 ± 4.4 vs -16.2 ± 6.0%, p = 0.015) or 6 segments (-9.0 ± 3.7 vs -14.6 ± 4.4%, p = 0.001), PC (0.86 ± 0.29 vs 1.32 ± 0.72 ml/mmHg, p = 0.01; PC/BSA (0.51 ± 0.17 vs 0.76 ± 0.41 ml/mmHg m,2 p = 0.013). Conclusion: Patients with ES have better RV function and less impaired PAS compared to patients with other types of PH and similar PVR. Moreover, besides RV function, PAS parameters emerged as predictors of cardiac death in non-ES patients that had worse prognosis than ES patients. The impact of these findings on clinical outcomes in ES patients remains to be further studied.