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The inclusion of pulmonary arterial pressure misclassifies diastolic function using the current EACVI guidelines in pre-capillary pulmonary hypertension

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Background: Pulmonary hypertension (PH) can be pre-capillary or post-capillary (PVH) etiology based on left-sided filling pressures and pulmonary vascular resistance. The 2016 EACVI/ASE Recommendations for the Evaluation of Left Ventricular Diastolic Function (LVDF) provides flow-diagrams to categorize patients. Parameters used include left atrial volume, Doppler-derived transmitral and mitral annular velocities, and systolic PA pressure (sPAP). There are no dedicated criteria to assess the diastolic function in pulmonary arterial hypertension (PAH). Additionally, diseases such as scleroderma can result in both PAH and PVH, thus including sPAP may alter LVDF diagnostic reliability in this population.

Purpose: Because elevated PAP is fundamental to PAH, we hypothesized that the EACVI/ASE diastolic function algorithm has a lower predictive value in correctly classifying diastolic function in scleroderma.

Methodology: We performed a single-center retrospective analysis of scleroderma patients who underwent complete echocardiography and comprehensive right and left heart catheterization for PH evaluation. PH categorization was defined using the 6th World Symposium hemodynamic definitions (PAH as mPAP ≥20 mmHg, PCWP ≤15 mmHg, PVR ≥ 3 WU). Diastolic function categorization used 2016 EACVI/ASE recommendations. Index catheterization and echocardiogram closest to cardiac catheterization were analyzed.

Results: 260 patients underwent evaluation and 63 were diagnosed with PH. PAH was diagnosed in 35 (age 64 ±10, mPAP 55± 18 mmHg, LVEF 60 ± 6%) and PVH in 28 (age 65 ± 10, mPAP 34 ± 14 mmHg, LVEF 63 ± 6%). Of the PAH patients, 20 had normal LVEDP (≤ 12 mmHg) and 15 increased LVEDP. In the PAH normal LVEDP patients, the EACVI algorithm classified diastolic function as normal in 25%, grade 2 in 5%, Grade 3 in 5%, and "indeterminate" in 65%. In the PAH group with increased LVEDP (> 12 mmHg), 27% were incorrectly identified as normal, 7% as grade 2 dysfunction, and 66% as indeterminate. The diastolic function algorithm has a sensitivity of 27% and specificity of 75% to diagnose a LVEDP ≤ 12 mmHg, with an AUC of 0.508 ($p = 0.91$). With exclusion of sPAP from the algorithm, indeterminate cases in both PAH groups were reclassified as normal, resulting in improved sensitivity (93%) but poorer specificity (10%), and a similar AUC (0.517, $p = 0.72$). In PVH patients, the algorithm performed better with a sensitivity of 63% and specificity of 83% to predict LVEDP > 12 mmHg with AUC 0.773, $p = 0.017$.

Conclusion: In scleroderma patients with PAH, the EACVI diastolic algorithm performs poorly and is confounded by including PAP as a parameter. The sensitivity of the algorithm is improved by the exclusion of sPAP although with reduced specificity. It remains inadequate to reliably diagnose normal LVEDP. While useful in other populations, algorithm modifications including exclusion of PAP, must be employed in suspected scleroderma PAH.