

Impact of Training Echocardiography in Connective Tissue Diseases

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DESCRIPTION

The Echocardiography describes the use of exercise testing to identify connective tissue disease (CTD) patients at high risk of developing pulmonary hypertension. The study discovers that people who had higher exercise-related increases in mean pulmonary artery pressure relative to cardiac output (mPAP/CO) were more likely to develop PH during the follow-up period. If confirmed in larger studies, this finding could lead to improved screening methods for patients with CTD (primarily scleroderma) and possibly other at-risk populations.

PH complicates many types of CTD, but it is most common in scleroderma and is the leading cause of death in this patient population. Annual PH screening is recommended in both scleroderma patients and those with mixed CTD with scleroderma features. The most common screening modality is echocardiography (at rest), which allows for the estimation of pulmonary artery systolic pressures as well as the evaluation of any right heart enlargement. When suspicious findings on an echocardiogram are detected, right heart catheterization (RHC) is recommended; however, a significant number of false negatives and false positives occur.

Using echocardiography in conjunction with pulmonary function testing, N-terminal pro-B-type natriuretic peptide levels, and other clinical results can improve screening accuracy. This approach, however, has several limitations. For starters, a negative screen only provides information about current PH risk, and despite a lack of research supporting this approach, annual testing is generally recommended.

Second, even with annual screening, some patients will be diagnosed with PH at an advanced stage, either as a result of a missed diagnosis during screening or as a result of rapid interval progression.

As a result, exercise testing is an appealing alternative. Previously, exercise echocardiography was used to detect "exercise-associated

PH," which was previously defined as an mPAP >30 mm Hg during exercise. Current PH guidelines advise against using this definition because some normal people will develop pulmonary pressure elevations in this range, usually as a result of achieving a cardiac output of more than 10 liters/min during exercise or from exercise-related increases in left atrial pressure.

Furthermore, the clinical significance of exercise-associated PH using this definition has not been established. Nonetheless, a growing body of literature in this field suggests that if the full pressure-flow relationship is taken into account, normal ranges and disease patterns can be defined. The current study has several limitations. Noninvasive estimates of PAP and CO can differ significantly from RHC values, especially during exercise, and the investigators do not directly compare their noninvasive pressure and flow measurements during exercise with invasively measured parameters. Furthermore, rapid drops in PAP and CO occur immediately after exercise cessation; thus, the chosen method may fail to identify patients with high mPAP/CO relationships at peak exercise, resulting in misclassification.

RHC was also only performed in individuals with an echocardiographic mPAP estimate of 25 mm Hg; thus, the sensitivity and specificity of their screening measure cannot be determined—though the relatively long follow-up (median 32 months) reduces the likelihood of PH being missed. The lack of evaluation for elevated left atrial pressure during exercise is also a limitation of the technique, as one would expect that heart failure with preserved ejection fraction would be common in PH in CTD (though not reported in this study) and would contribute to exercise-associated increases in mPAP.

Finally, while physiological principles suggest that post-exercise mPAP alone would not be as effective as mPAP/Q, in this cohort, mPAP at peak exercise was a strong predictor of future PH in the univariate evaluation, and evidence that mPAP/Q performed better than post-exercise mPAP would have significantly strengthened the study.

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