Prevalence and Predictors of Pulmonary Artery Hypertension is Systemic Sclerosis - Comment on the Article by Uma Kumar et al

Sir,

We read with interest the article by Uma Kumar et al regarding non-invasive screening of patients with Systemic Sclerosis (SSc) for pulmonary artery hypertension (PAH). Few joints warrant for explanation by the authors.

The first point is regarding Doppler echocardiogram as a tool for screening patients of SSc associated with a high risk of developing PAH. Uma et al considered the patients of SSc having PAH if they fulfilled either of two criteria- (a) Transtricuspid gradient > 35 mmHg (b) Right ventricular acceleration time < 90 milliseconds. Tricuspid regurgitation was present in 45% of their patients and 18% had a transtricuspid gradient of > 35 mmHg. In this context the echocardiographer’s experience is crucial for accurate ultrasonographic identification of tricuspid regurgitation and hence determination of PAH. In the previous study from France involving 709 patients, velocity into isolated and secondary form based on the presence or absence of interstitial lung diseases on HRCT chest. PFT results were available in all 100 patients but diffusing lung capacity (DLco) was done only in 76 patients. Also, the autoantibody profile was not evaluated in detail.

Additional screening tests are needed in order to identify early forms of PAH. Pro-BNP is a biomarker that correlates well with hemodynamic measures and predicts survival in patients with PAH associated with SSc. The specificity and sensitivity of Pro-BNP to predict PAH are similar to those of echocardiography.

The second point is regarding classification of patients. In the study by Uma et al an attempt was made to classify PAH into isolated and secondary form based on the presence or absence of interstitial lung diseases on HRCT chest. PFT results were available in all 100 patients but diffusing lung capacity (DLco) was done only in 76 patients. Also, the autoantibody profile was not evaluated in detail.

It has been proposed that several predictive factors including disease subset (limited or diffuse), autoantibodies, and pulmonary function tests can help to classify patients into 3 groups:

1) Isolated PAH have a long history of Raynaud’s phenomenon, limited scleroderma, anticientromere antibody positivity, an extremely low diffusing capacity for carbon monoxide (DLco), near-normal forced vital capacity (FVC), minimal pulmonary fibrosis, and an FVC:DLco ratio of > 1.8 at the time of the diagnosis.

2) Patients with diffuse systemic sclerosis and pulmonary interstitial fibrosis have antitopoisomerase antibody (SCL-70 antibody) and a very low FVC and a DLco that is decreased to a similar degree, so the FVC:DLco ratio remains close to 1.

3) Patients with mixed ILD-PAH have antinucleolar antibody, moderate interstitial fibrosis and then later develop severe PAH out of proportion to the degree of fibrosis. The patients have vasculopathy in addition to the fibrosis. The FVC:DLco ratio is > 1.8. Recently a multicenter study is being conducted by members of the Scleroderma Clinical Trials Consortium (Pulmonary Hypertension Assessment Registry of Scleroderma - PHAROS). The consortium’s website is www.sctc-online.org. PHAROS participation will give best way to contribute to really making a difference in scleroderma.

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Reply from Author

Sir,

We thank Dr. Chogle and his colleague for their interest in our study "Prevalence and predictors of pulmonary artery hypertension (PAH) in systemic sclerosis (SSc)". We fully agree with the view that detecting tricuspid regurgitant jet is operator dependent. It can be assessed in 39%-86% of patients, and careful Doppler examination by experienced sonographer can detect tricuspid regurgitant signals in 74% patients. In our study Doppler was done by a consultant cardiologist with considerable experience and tricuspid regurgitant jet was found in 45% patients.

Regarding their second point, role of Pro-BNP in SSc-PAH, recently a few studies have highlighted its role in PAH but it is still not recommended as screening modality for PAH in...
SSc. Also, there is paucity of large population based studies on the status of Pro-BNP in patients with systemic sclerosis without known PAH.

We could not do diffusing lung capacity (DLco) in all patients because of technical reasons. While a complete autoantibody profile would have been ideal, it was not the primary objective of the study and was not done due to resource constraints.

We do hope that are outside, as do the pertinent comments of Chogle and Shah, draw attention to this often overlooked problem in SSc.

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