Author reply to the editor

Sir,
I read with great pleasure the comments from Dr. Majdy Idrees on our recent article in Annals of Thoracic Medicine[1] and am willing to respond to him.

Pulmonary hypertension (PH) can complicate the course of many forms of interstitial lung disease (ILD). However, diagnosing PH in the setting of ILD can be very challenging, as the manifestations are subtle and the two conditions commonly share similar symptoms. When PH was suspected in our study, we performed a thorough screening that included clinical examination, assessment of physiological parameters, computed tomography and/or echocardiography. Since these

non-invasive tests failed to prove reliable for predicting PH in the context of ILD, however, right heart catheterization (RHC) remains the reference standard for the diagnosis of PH. This invasive procedure it is considered extremely safe when performed in an experienced center. None of the patients in our study developed any complication after RHC and the safety of this procedure was previously well-documented in a large cohort of patients (n = 7218) with PH due to various etiologies who underwent RHC and showed an overall procedure-related mortality of 0.05%.[2] Despite the good safety profile of RHC, however, the procedure should only be performed by experienced clinicians. Because we currently lack noninvasive methods for accurately detecting PH in the context of ILD and do not yet have consensus guidelines for recommending RHC in this setting, RHC should only be performed in a reference center with significant interest and expertise in many forms of ILD.

In his correspondence, Dr. Idrees correctly stated that pulmonary arterial hypertension (PAH) targeted therapy has not been approved for the treatment of non-PAH groups, including PH groups III and V. As we noted in our paper, only a subset of patients in groups III and V received PAH targeted therapy. Among the treated participants, sildenafil therapy was given to patients with idiopathic pulmonary fibrosis (IPF) and chronic hypersensitivity pneumonitis and sildenafil, bosentan and nebulized iloprost were offered to those with connective tissue disease-associated ILD and sarcoidosis, either as monotherapies or in combination, as clinically indicated. The fraction of group III and V patients who received PAH-specific therapy in our study was similar to those in the PH registry recently reported from the UK.[3] In our center, we use PAH-specific therapy in group III and V patients on a case-by-case basis. Our patients are fully aware of the lack of supporting data and the potential risks of such therapy. Moreover, once a specific therapy is started, we monitor our patients very carefully for objective and subjective evidence of improvement. If these goals are not met, we withdraw PAH therapies.

Although no therapy has yet been proven to alter the outcome in patients with PH-associated ILD, clinicians should dedicate their efforts to improving the quality-of- life and functional capacity of these patients. In support of this notion, when IPF patients with evidence of right ventricular dysfunction were treated with sildenafil, improvement in quality-of-life and preservation of exercise capacity were noted.[4] A similar finding was also obtained among sarcoidosis-associated PH patients treated with PAH-specific therapy either as a monotherapy or in combination.[5-7]

Clinical trials that specifically address the efficacy of PAH-targeted therapy among groups III and V are greatly needed and in the absence of a prospective national PH registry, these patients should only be investigated and treated in an appropriate reference center.

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