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Authors' Reply

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We read with genuine interest the correspondence of Deepak Aggarwal and Phiza Aggarwal, in which they commented on our article demonstrating the prognostic value of pulmonary hypertension (PH) in initial evaluation of idiopathic pulmonary fibrosis (IPF) [1].

We reported that mean pulmonary artery pressure (MPAP) at the initial evaluation was an independent determinant of survival along with forced vital capacity in IPF. In this study, multivariate analysis demonstrated that arterial oxygen saturation during exercise, diffusion capacity to carbon monoxide, and dyspnea score had no significant impact on survival, but forced vital capacity and MPAP did. On this point, we argued the importance of measuring MPAP as it may provide useful information in considering possible treatments [2]. Admitting that it is not completely noninvasive, we consider that right heart catheterization is safe with few complications when done in experienced institutions [3]. Estimation of pulmonary artery pressure by echocardiography is reported to be often inaccurate [4].

In terms of patients enrolled, our study also included patients without pathological diagnosis. We suppose this may better reflect real clinical settings, as the new IPF guideline supports clinical diagnosis of IPF without pathological confirmation in cases with the presence of a usual interstitial pneumonia pattern on high resolution CT findings [5]. Additionally, a comprehensive serological analysis and detailed physical examination of connective tissue diseases were performed in our study, so we are confident of the diagnosis of IPF in our study.

As for treatments in IPF, many clinical trials have had negative outcomes with the notable exceptions being the pirfenidone trials conducted in Japan [6] and the rest of the world, and a first major step forward has been taken with the Japanese and European approval of pirfenidone for patients with IPF [7]. The earlier the diagnosis is made, the sooner therapy can be initiated, which, in turn,

may reduce the decline in lung function and improve the overall prognosis [8].

Although the importance of early detection and management of pulmonary arterial hypertension is well recognized, it is still unclear whether that is also true for PH associated with IPF. However, considering the poorer prognosis in patients with even marginal PH (MPAP 21–25 mm Hg), the importance of earlier diagnosis of PH associated with IPF should be intensively investigated. Without question, further clinical trials including new agents and drugs targeting pathways implicated in the pathogenesis of pulmonary fibrosis and PH are needed to improve the overall outcome of IPF [9].

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