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Is Pulmonary Hypertension Without Left Ventricular Dysfunction Associated With Poor Survival in Patients With Sarcoidosis or Is It Just a Factor of Advanced Disease Stage?

To the Editor:

We read with great interest the article by Baughman and colleagues1 in a recent issue of CHEST (November 2010). The study concluded that pulmonary hypertension (PH) without left ventricular dysfunction (LVD) was associated with increased mortality in a population sample of patients with sarcoidosis. Although we strongly agree that patients with PH need to have right-sided heart catheterization for better classification and hence treatment strategy of PH, we have some reservations about the study.

The difference in the two main study groups, namely PH without LVD (PH/no LVD) and PH with LVD (PH/LVD) was barely significant, with a CI almost touching the null value. On the survival curve (Fig 4 in the study1) at 3 years, the CIs of the two groups were insignificant and overlapped (at the time when 23 of the 50 PH/no LVD and only 11 of the 20 PH/LVD were alive). This indicated that with a possibly severe PH stage and early death (within 3 years) the difference in mortality in the two groups was insignificant. The fact that increased pulmonary vascular resistance in the same study was an independent predictor of mortality regardless of the cause of PH may support this observation.

The study attempted to firmly stratify the relationship between the cause of PH in patients with sarcoidosis and increased mortality, with some adjustment in statistical analysis using Cox proportional hazards regression for important and known factors that are associated with increased mortality in this population, such as reduced diffusing capacity of the lung for carbon monoxide and home oxygen use. Other critical factors, such as 6-min walk distance and whether patients received treatment and the type of treatment, were not known or controlled for in the study population because of the retrospective nature of this study.

The question remains: Is PH without LVD the most important factor in predicting survival, or is it only one factor among many others indicating increased sarcoidosis disease severity and lung parenchymal destruction? In a future study, it might be of interest to calculate a score based on the diffusing capacity of the lung for carbon monoxide, FVC, home oxygen use, radiographic stage, PH without LVD, and 6-min walk distance and to test the relationship of these individual factors, as well as the total score, on survival. Many of those factors were not associated with increased mortality in this study but were significant factors in other studies.3-4

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Response

To the Editor:

We appreciate the comments of Drs Alrajab and Abu Baker about our recent article in CHEST (November 2010). The question they raise about whether pulmonary hypertension (PH) without left ventricular dysfunction (LVD) was significantly different in survival rates from PH and LVD is well taken. We found that the survival rates were different for patients with PH, with those patients without LVD having worse survival rates. The differences were most apparent with longer follow-up periods, and this may be because of the inclusion of patients with PH and LVD who had cardiacopathy. We did acknowledge this in our article and took pains to try to clarify that the differences were due to hemodynamics rather than associated factors. We agree that other factors, including treatment, may influence...