Outcomes in Patients with Idiopathic Pulmonary Fibrosis and Pulmonary Hypertension

This poster entitled Outcomes in Patients with Idiopathic Pulmonary Fibrosis and Pulmonary Hypertension was a single-center, retrospective study presented by Collins and colleagues. They presented 71 patients with idiopathic pulmonary fibrosis that also had right-heart catheterization data. Of those, 13 did not have pulmonary hypertension at all. And the rest were split between precapillary, postcapillary, and mixed pulmonary hypertension. There really was no difference between these groups as far as lung function goes, as measured by forced vital capacity, or FEV1. The diffusion capacity was not presented in this poster.

And there was no difference between these groups in their transplant free survival. This was possibly related to the fact that this was a small cohort as we do know that pulmonary hypertension increases the risk of mortality in patients with idiopathic pulmonary fibrosis from other cohorts.

This study did show that there was a mix of patients with precapillary, postcapillary, and mixed pulmonary hypertension. And this could have implications as far as management goes. We can't assume that all patients with pulmonary hypertension and idiopathic pulmonary fibrosis are really the same.

This is a retrospective review on outcomes in patients with idiopathic pulmonary fibrosis and pulmonary hypertension done by Ashley Collins and colleagues. This was a fairly well done retrospective review of 750 patients with idiopathic pulmonary fibrosis. They reviewed this and found patients who had validated right-heart catheterizations, and look for outcomes of transplant-free survival is the primary measure comparing patients with pulmonary hypertension in IPF, and those without.

They looked at five main groups, including those was precapillary pulmonary hypertension, postcapillary hypertension, uncategorized pulmonary hypertension, mixed pulmonary hypertension, and those with right-heart catheterizations with no evidence of pulmonary hypertension. There were 29 patients with precapillary pulmonary hypertension, and then a combination of others that made up the 71 patients with data.

And basically, they found that in aggregate, there was no difference in transplant-free survival among these different groups of subsets of pulmonary hypertension in patients with idiopathic pulmonary hypertension, but that in total, patients with pulmonary hypertension compared to those without have worse outcomes and less transplant-free survival time compared to people with IPF who don't have PH.

And the key takeaways from this are that, more work is needed to characterize patients with types of pulmonary hypertension with IPF, and better understand prevalence of these subsets of disease and to understand impacts it has, as a disease process, in patients with IPF on prevention, and co-morbidities, and survival time. So I think that it was a very large body of patients that were evaluated and characterized as pulmonary hypertension and its relationship to survival in this patient population.

So some of the clinical implications in practical application of this poster would indicate that practicing clinicians should refer patients with pulmonary hypertension and worsening oxygen status for early right-heart catheterization. They should be aware that the presence of pulmonary hypertension adds significant morbidity and mortality to patients with IPF, and that defining the process that is underlying their pulmonary hypertension may have substantial clinical relevance as further research becomes available.