Characteristics of Patients in the IPF-PRO Registry

This presentation by Dr. Culver on the Idiopathic Pulmonary Fibrosis Prospective Outcomes Registry describe the baseline characteristics of this cohort. Two-thirds of these patients had definite UIP on their high resolution CT imaging, and about one-seventh of them had evidence of concomitant emphysema. This cohort was open to all commerce, including those with mild to very severe idiopathic pulmonary fibrosis. What they found in this baseline study was that patients had a bit more severe disease than those in some of the previous clinical trials that have been published. They also had impaired quality of life as measured by multiple health-related quality of life questionnaires. There was a high prevalence of comorbid conditions. Most prevalent was gastroesophageal reflux disease, coronary artery disease, and obstructive sleep apnea. These patients were also on multiple other medications for their comorbid conditions, including statins and proton pump inhibitors being the most common. More than 50% of them were on antifibrotics at the time of enrollment, and 12% of them, very few, were on oral steroids at the time of enrollment.

The take-home points of this presentation on the IPSPRO Registry are that at baseline the patients enrolled into the registry were mostly elderly, white, and male with impaired lung function, impaired health-related quality of life, and many comorbid conditions and other medications that they took. There is a plan for longitudinal data collection from this registry that will help us to inform our knowledge of idiopathic pulmonary fibrosis and its behavior over time.

This was a fairly well done descriptive study of patients with idiopathic pulmonary fibrosis by Kim and colleagues describing patients all admitted to hospitals with IPF for various reasons, including but not limited to IPF exacerbations. They found that this cohort meets similar demographic characteristics in that most of the patients were older with a median age of around 70. The majority of the patients were Caucasian. The majority were male. They had a median FEC of around 70%. Only 20% of the patients ended up getting bronchoscopy as part of their standard of care. About two-thirds had antibiotic treatment, and around half were treated with IB steroids empirically most of the time.

You can tell there's a dichotomy from this and that the patients who were not requiring vent support had a much higher discharge home rate, and patients with IPF that required ventilatory support in hospital had a higher mortality rate, which is described in similar studies and known that patients have substantial risk of death in hospital on mechanical ventilation with IPF.

Some of the main takeaways from this are that patients with IPF being admitted to the hospital, if it's for a respiratory condition, bronchoscopy is rarely used in the treatment of this, may be controversial and high risk for bronchoscopy and patients that could be implicated. Patients requiring mechanical ventilation may require early consultation with palliative care as they have a high mortality rate in general.