The Landscape of Interstitial Lung Disease from Real-World Data – Including Anti-Fibrotic Drugs

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Interstitial lung diseases (ILDs) represent a heterogeneous group of progressive disorders with limited treatment options and poor prognosis. While randomized controlled trials (RCTs) remain the gold standard for evaluating therapeutic efficacy, their strict inclusion criteria often limit generalizability to real-world patient populations. In contrast, real-world data (RWD) can capture diverse patient backgrounds and treatment practices, providing complementary evidence to bridge this gap. In this lecture, I would like to provide an in-depth exploration of the real-world landscape of ILDs, with a primary focus on idiopathic pulmonary fibrosis (IPF), using data from our cohorts and the National Database of Japan (NDB Japan), the world's largest claim-based database. Analyses of NDB Japan demonstrate that the prevalence and incidence of ILDs and IPF have increased over time in Japan, a trend consistent with observations in other countries. When examining the impact of antifibrotic drugs, we identified several clinical significant impacts. To date, a survival benefit of antifibrotic therapy in IPF has not been fully established, as survival data are subject to major biases such as immortal time bias. We carefully analyzed the survival effects of antifibrotic drugs while minimizing immortal time bias, and our findings indicated its potential survival benefit. Furthermore, antifibrotic drugs may also reduce the incidence of acute exacerbations, potentially altering the causes of death in IPF patients. Although it is well known that IPF patients are at higher risk of developing lung cancer than the general population, data from our cohorts as well as from the NDB Japan showed that antifibrotic therapy reduced the cumulative incidence of lung cancer in IPF patients. Interestingly, treatment with nintedanib, but not pirfenidone, was associated with a reduced risk of other cancers beyond lung cancer. In addition, we found that the use of hypnotics was associated with poorer prognosis, and that the prognosis of COVID-19 patients with underlying ILD was particularly poor. Taken together, these RWD provide a more comprehensive understanding of ILD, and contribute to improvements in our clinical practice.