PRECISION-ILD: PRECISION-ILD

Description

Interstitial Lung Diseases (ILDs) are a heterogeneous group of >100 different rare diseases, which share the fate of progressive scarring and, ultimately, death. Two anti-fibrotic drugs have demonstrated to slow-down fibrotic progression and steroids/immunosuppressants are commonly used for inflammatory-driven ILDs. However, patient's response to therapeutic options is variable and unpredictable. Similarly, setting a correct diagnosis is difficult in most cases, especially when patients are too sick for invasive procedures.

To investigate the differences and commonalities in genetic, genomic and environmental exposures/lifestyle in fibrotic ILDs depending on the entity, disease behavior (progressive fibrosis) and treatment response; (2) To integrate the biomarkers that most impact on prognosis and treatment response in diagnostic algorithms; and (3) To explore the feasibility and cost of implementing a P4 strategy in clinical practice for fibrotic ILDs.

Methods: We will extend, update and unify existing ILD cohorts (Spanish SEPAR ILD Reg, Observatory IPF.cat, CIBERES IPF and Familial ILD cohorts) in whom we will: (1) record demographic, epidemiological, clinical, physiological and lung morphology (radiological +/- histological) information; (2) obtain genetic variation, telomere length, and serum protein markers; (3) investigate environmental exposures (including air-pollution), (4) apply to integrative analytical methods to identify endotypes, predictive biomarkers of disease trajectories, theragnostic biomarkers and new therapeutic targets. Results (5) will be validated in other fibrotic ILD cohorts (e.g.EuILDRegistry, Mexican fibrotic ILD Registry). Besides, we will explore how to translate this P4 medicine approach in clinical practice; (6) implementing a predictive score for prognosis and improving the diagnostic approach through biological data to reduce invasive procedures, and (7) estimate educational requirement and potential health cost implications.

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