

TITLE: Redefining Idiopathic Interstitial pneumonias with Genomics

INVESTIGATORS: Kevin K Brown & Roland M du Bois

ABSTRACT

There is an emerging consensus, at least for several neoplasms, that gene expression profiles (transcriptomes) can identify new clinically meaningful subsets of disease and predict clinical outcomes. The idiopathic interstitial pneumonias (IIPs) comprise a group of disorders of unknown etiology that result in diffuse pulmonary fibrosis. Considerable overlap exists across individual members of this group of diseases and outcome varies considerably within each constitutive member. It is likely therefore that current classification remains imprecise and that each individual disorder under the present classification actually comprises a heterogeneous group of entities. To better define this group of diseases we hypothesize that molecular characterization using gene expression profiles, taken in the context of traditional indices, will allow further disease sub classifications to be defined. To pursue this challenge, we hypothesize that the IIP transcriptome is controlled by genetic variants in the genome and that, in aggregate, these molecular features can be used to redefine this disease. To test this hypothesis, we will combine the results of genome-wide RNA expression with regions of interest from gene mapping studies to develop and validate a phenotypically anchored molecular signature for IIP and its constituent diseases.