Reviewer’s report

Title: Incorporating genetics into the identification and treatment of Idiopathic Pulmonary Fibrosis

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Reviewer: Paul Wolters

Reviewer’s report:

This is a very well written manuscript summarizing the importance of recent reports of genetic variants associated with IPF. The manuscript will be useful to readers because it relates the findings to important clinical scenarios and foreshadows future studies needed to better understand the genetics of lung fibrosis.

The MS could be improved by clarifying several minor points:

Although the authors comment that “the genotyping of patients is not a generally accepted strategy”, for evaluating IPF patients, could it useful in some circumstances? For example, is genotyping for TERT/TERC mutations useful in patients with a syndrome suggestive of a telomeropathy (early greying of hair, lung fibrosis, macrocytosis, etc) being considered for lung transplantation? The purpose would be that if the mutations were present, the patient may be at risk for post-transplant cytopenias. Knowing their genotype could assist with patient management.

Risk for family members section: The authors comment regarding patients in the 1986 Bitterman study “….whether these individuals progressed to development of pulmonary fibrosis themselves was not studied” is incorrect. El-Chemaly et al reported 27-year follow-up for two of the patients in a 2011 publication in Chest. Both of the individuals developed pulmonary fibrosis. Although the sample size is limited, this report should be cited in the MS.

Page 7 of document: “The association of the MUC5B promoter polymorphism appears to be specific to pulmonary fibrosis”. Should probably read “….. specific to idiopathic pulmonary fibrosis”.

Figure 1. Listing TERT and TERC as both very rare and common variants is confusing and contradictory. Are they rare or common?

Quality of written English: Acceptable

Statistical review: No, the manuscript does not need to be seen by a statistician.

Declaration of competing interests:
No competing interests