Reviewer's report

Title: Preliminary Results from the Finnish IPF Registry

Version: 2  Date: 26 April 2015

Reviewer: MARIA MOLINA-MOLINA

Reviewer's report:

Kaunisto J et al present data of the Finnish IPF Registry and demonstrate that the expert evaluation of ILD patients is crucial for obtaining accurate epidemiological data and to make decisions about the treatment indication. Currently, to properly estimate prevalence and incidence of IPF is important for the health worldwide national care systems since a new era of IPF treatment has been opened, starting with Pirfenidone and Nintedanib (orphan drugs for a rare disease but with a high cost). So, the present study is relevant, mainly because two points; 1) The role of re-evaluation under an expert ILD team, 2) to describe the main features of IPF cases in a multidisciplinary and multicentre national approach. The methods are appropriate and well described. However, there are major concerns that should be discussed to improve the quality of the paper (results-discussion-conclusions) and some minor concerns. The writing of the report is acceptable.

Major concerns;

1. The authors talk about “early or relatively early stage” of IPF just because the FVC-DLCO mean values. However, 14 patients died before the end of 2012, 76% of cases showed honeycombing on HRCT and typical UIP in 87%, and, finally, the time from the onset of symptoms to the date of diagnosis was 1.9 years. The patients were classified in mild-moderate stage, but it is argued if most patients were diagnosed in an “early” stage. This term is incorrect for this cohort, the authors can refer mild-moderate stage but not “early”. The last sentence of Conclusions should be changed by “mild-moderate stage”.

2. If 87% of cases presented a typical UIP radiological pattern, how only 76% showed honeycombing?

3. 70-80% were not really IPF, the authors should present more in detail how these cases were diagnosed as another ILD. What was the cause of the overdiagnosis of IPF in this cohort? (radiological and clinical mistakes in non-expert physicians).

4. Patient data was re-evaluated by pulmonary physicians and radiologists in order to estimate the prevalence of IPF, what about the pathologists for the 27 cases with lung biopsy? The role of the pathologist is crucial in some IPF diagnoses and it seems that the authors did not include any pathologist in the ILD multidisciplinary team for the evaluation, why? If they did it, they should include it in more detail in the Results. Only in discussion the authors mention the possible role of pathologists.
5. Another limitation of the estimated IPF prevalence, not only in this cohort but also in other Registries based on ILD codes, is that some ILD cases could be misdiagnosed and, therefore, not codified. It should be included in discussion.

6. If there is no data about IPF treatments in this cohort why the authors include the last sentences in Discussion about Nintedanib or Pirfenidone? The authors should include data about IPF treatment in the cohort or to delete at least the last sentence (to avoid unrelated speculations).

7. Smoking history was different among different centres. There is no data about the possible relationship of this difference with the outcome (relationship with FVC-DLCO values? Reported deaths?

8. 14 patients died during 2012, could you include more clinical details about these cases?

Minor concerns;
- 10 from 18 cases with possible UIP radiological pattern were still included without lung biopsy, could you explain better why? (because follow-up? Family history?...).
- The sentence of Discussion about study limitations; “Only a small percentage of patients were excluded after the re-evaluation of the diagnostic HRCTs, which in part proves the high quality of the public care health system”. It would be more correct if it points; “…the high quality of the radiological evaluation of the public health care system…”, since previously a high % of cases were discharged as not IPF and this sentence refers specifically to the HRCTs evaluation.

Level of interest: An article of importance in its field

Quality of written English: Acceptable

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