Disease progression in IPF assessed using pulmonary function tests and functional respiratory imaging (FRI) – a pilot study

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Hypothesis

Disease progression is correlated with a reduced lobar volume, an enlarged airway volume and a reduced airway impedance.

Methods

In this trial we studied 5 IPF patients for 6 months to assess the changes in airway structure and function. At the baseline and 6 month visit FVC, DLCO and helium based lung volumes were measured. In addition, HRCT-based functional respiratory imaging (FRI) was performed in all patients. FRI provides information regarding lobar volumes, regional airway volumes and impedance.

Results and interpretation

In terms of FVC, 1 patient declined (-6%), 1 patient improved (+6%) and 3 patients remained stable (-1%,0%,1%). The patient that declined in FVC also experienced a -21% in DLCO, -12.67% in FRC and -7.47% in TLC. While the airway volumes (iVaw) stayed constant, the airway resistance (iRaw) decreased by -6.7%. However when the values are made specific by correcting for the lung volume (siVaw = iVaw/lung volume, siRaw = iRaw * lung volume) then the specific image-based airway volumes (siVaw) in this patient showed an increase
in airway of 23.99% and the specific airway resistance reduced by 30.90%. This finding is in line with the hypothesis. When correlating the change in FVC with the change in \(iVaw\) for all patients, a negative but not significant correlation (\(R = -0.6, p = 0.28\)) was found. However a significant inverse correlation was found for all patients between the change in FVC and the change in \(siVaw\) (\(R = -1, p = 0\)); and changes in lobar volume and the \(siVaw\) in the upper lobes (\(R = -0.71, p = 0.00283\)). Furthermore it was found that lobes with a larger increase in \(siVaw\) attract a larger fraction of the incoming air (\(R = 0.42, p = 0.035\)), thereby suggesting that the effect of increased in airway volume is larger than the effect of increased impedance.

**Conclusions**

By PFT criteria, the disease only progressed in one patient, but the study suggests that disease progression is associated with a reduction in regional lung volume, an increase in regional airway volume and a reduction in regional impedance. Further studies need to confirm these initial observations.