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

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Rationale
Disease progression in IPF is correlated with reduced lobar volume (iLobe), enlarged airway volume (iVaw) and reduced airway impedance (iRaw).

Methods
5 IPF patients were followed for 6 months to assess changes in airway structure and function. At baseline and after 6m FVC, DLCO and 3He-based FRC/TLC were measured. Also, HRCT-based functional respiratory imaging (FRI) was performed. FRI provides information on iLobe, iVaw and iRaw.

Results
In terms of FVC, 1pt declined (-6%), 1pt improved (+6%) and 3pts remained stable (-1%,0%,1%). The pt that declined in FVC also experienced: DLCO -21%, FRC -12.67%, TLC -7.47%. iVaw stayed constant, iRaw decreased by 6.7%. However when iVaw/iRaw are corrected for lung volume (siVaw = iVaw/iLobe, siRaw = iRaw*iLobe), then siVaw increased by 23.99% and siRaw reduced by 30.90%. A negative but not significant correlation was found between changes in FVC and iVaw (R= -0.6, p=0.28). However, significant correlations were found between changes in FVC and siVaw (R= -1, p=0); changes in iLobe and siVaw (in the upper lobes R= -0.71, p=0.003); and changes in siVaw and lobar ventilation (R=0.42, p=0.035).
Conclusions

By PFT criteria, IPF only progressed in one patient, but the study suggests that IPF progression is associated with a reduction in iLobe, an increase siVaw and a reduction in siRaw. Further studies need to confirm these initial observations.

Figure

Figure 1 Regional changes in lobe volume for 1 patient in the trial