European Respiratory Society
Annual Congress 2012

Abstract Number: 2268

Publication Number: P1450

Abstract Group: 7.3. Cystic Fibrosis

Keyword 1: Imaging **Keyword 2:** Longitudinal study **Keyword 3:** Monitoring

Title: Tracking disease progression in cystic fibrosis using bronchiectasis, trapped air and quality of life

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Body: Background. Progression of cystic fibrosis (CF) is characterized by bronchiectasis (BE) and trapped air (TA) on CT. We hypothesize that progression of BE, and TA results in a lower Health Related Quality of Life (HRQoL), assessed by the Cystic Fibrosis Questionnaire-Revised (CFQ-R). Objective. To evaluate associations between changes in BE, TA, and CFQ-R over time. Methods. Cohort study (July 2007-January 2012). Clinical stable CF patients, with two routine bi-annual chest CTs, and CFQ-Rs. CT scans were anonymous and randomly scored, using CF-CT BE and TA scores. Scores are expressed as % of maximum score. CFQ-R was completed by children (aged 6-13 years) and adolescents (aged ≥ 14 years). Score-range 0-100, higher scores indicate better HRQoL. For changes in CF-CT BE, TA score, CFQ-R respiratory-symptoms domain scores, their correlations, and to test the hypothesis we used Student's paired t-test, Spearman's correlation coefficient, and linear regression model, adjusted for age and gender. Results. CF patients (n= 40): mean age T_1 11.9 years, T_2 13.8 years. In two years there was a significant increase in CF-CT BE scores (p=0.03) and CF-CT TA scores (p=0.03), but not in CFQ-R scores. At T_1 CF-CT BE scores (p<0.01, r=-0.49) and CF-CT TA scores (p=0.04, r=-0.34) correlate with CFQ-R scores. At T_2 similar correlations were found: CF-CT BE (p<0.01, r=-0.41), CF-CT TA (p=0.02, r=-0.37). Change in CF-CT BE and TA scores did not correlate to change in CFQ-R scores. Conclusion. BE and TA correlates to HRQoL. Modest progression of BE and TA did not result in impairment of HRQoL. Supported by an unconditional grant by Gilead Sciences, Inc and the Sophia fund: “steun door Zeevaart.”