**99 Early predictors of bronchiectasis and trapped air severity in cystic fibrosis**

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**Objective**: Bronchiectasis (BE) and trapped air (TA) are important determinants of cystic fibrosis (CF) lung disease. We aimed to identify early predictors of BE and TA severity in children with CF.

**Methods**: Retrospective analysis. Inclusion: Availability of sweat chloride at time of diagnosis and of at least one routine in- and expiratory spirometry-controlled bi-annual volumetric CTs, at least 5 years apart, aged 6–13 years. Exclusion criteria: severe lung disease assessed by chest computed tomography (CT) at CT1, pulmonary exacerbations (iv antibiotic courses in >7 days), pancreatic exocrine insufficiency at CT1, or very easy. As reflected in total treatment and inhalation time, and number of breaths. All CF patients could handle the device properly and most of them rated its use as easy or very easy.

**Results**: Twenty-seven patients were studied, median age 9.2 (6.9–12.9) years, median time interval between CTs 6.5 (5.8–7.9) years. Children with Pseudomonas over had 7.91 (95% confidence interval 2.26–13.58) % higher BE score at CT2 (p < 0.01). An exacerbation prior to CT1 resulted in a 22.8 (1.64–43.2) % higher TA score at CT2 (p < 0.01). A 1% increase on BE score at CT1 resulted in a 0.04 (0.67–1.41) % higher BE score at CT2 (p < 0.01). A 1% increase on BE score at CT1 resulted in a 1.57 (0.06–3.08) % higher TA score at CT2 (p < 0.04). Multivariate analysis showed that the BE score on CT1 remained predictive for BE (p < 0.01) and TA (p = 0.04) at CT2.

**Conclusion**: Pseudomonas or exacerbation prior to CT1 predicted BE and TA at CT2 (median) 6.5 years later. This effect appeared to be mediated through BE at CT1, which was predictive of BE and TA severity 6.5 years later.

**Funded by Gilead Sciences, Inc and the Sophia fund “Steu door zeevaart”**

**200 Is sweat chloride predictive for severity of cystic fibrosis (CF) lung disease assessed by chest computed tomography (CT)?**

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**Background**: CF lung disease is characterized by poor genotype-phenotype correlation. Sweat chloride level is a functional marker of the CF Transmembrane Regulator protein and could be a predictor of the severity of CF lung disease.

**Objective**: To study correlations between sweat chloride and bronchiectasis (BE), trapped air (TA) and mucus plugging on chest CT.

**Methods**: Retrospective analysis. Inclusion: Availability of sweat chloride at time of diagnosis and of at least one routine in- and expiratory spirometry-controlled volumetric chest CT scan. CT scans were de-identified and randomized (Myrianb Montpelier, Fr) and scored using the CF-CT scoring system (%max). Intra- and interobserver variability: intraclass correlation coefficient. Associations between sweat chloride-levels and CF-CT subscores: Pearson’s correlation coefficient and multivariate regression models. Effect modification age CT-scan tested by stratification. Descriptives: expressed as median (range).

**Results**: 69 Children (34 male), age sweat chloride 0.8 (0–19.5) years, age chest CT 13.8 (5.5–19) years, TA score 6.0 (0–25.3) %max and BE score 2.0 (0–24.3) %max. Univariate analysis: CF-CT BE scores vs sweat chloride (μ). Multivariate models adjusting for age of sweat test and CT scan: significant association between sweat chloride and CF-CT BE (p = 0.036) and mucus plugging (p = 0.027). Stratification in terciles for age of CT scan showed that association was present only in the oldest age group (range 15–19 years).

**Conclusion**: Sweat chloride levels are predictive of long term CF lung disease as determined by chest CT, the association was primarily determined by children older than 15 years.

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**198 Time and handling of the I-Neb in target inhalation mode (TIM) and tidal breathing mode (TBM) assessed in mild to severe CF patients**

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**Objectives**: For effective treatment of CF with inhalated drugs, efficient, easy to use devices are needed. This study was designed to assess time and practicability of Alpha-1-Antitrypsin (A1-PI(H)), (77 mg/1.1 ml) lung deposition in CF patients.

**Methods**: Using the Respironics I-Neb AAD System. 15 mild to severe CF patients (FEV1: 34–101%) were included to inhale study drug in two inhalation modes: Target Inhalation Mode (TIM) and Tidal Breathing Mode (TBM).

**Conclusion**: Total inhalation time was lower in TIM (3±0.5 min) than in TBM (4.5±2.1 min). Inhalation time and volume inhaled per breath were lower in TIM than in TIM. Subjects inhaled more deeply and slowly in TIM. Consequently, treatment time was longer and the number of breaths needed to deliver a similar dose was higher in TIM than in TBM. In both inhalation modes a high percentage of CF subjects rated their mouthpiece exhalation as “easy” and “very easy” (TIM: 80%, TBM: 100%). In total, 87% of the CF subjects classified their mouthpiece inhalation through in TIM as “easy” and “very easy” (TIM: 40%). For both inhalation modes a high percentage of CF subjects classified the use of the I-neb as “easy” and “very easy” (TIM: 71.5%, TBM: 86.7%).

**All CF patients were able to inhale the full dose in <6 min in TIM and <10 min in TBM. TIM is consistently more efficient and less time consuming than TBM, as reflected in total treatment and inhalation time, and number of breaths. All CF patients could handle the device properly and most of them rated its use as easy or very easy.**