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Title: Multiple breath washout for monitoring alpha-1-antitrypsin deficiency related lung disease

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Body: Patients with alpha-1-antitrypsin deficiency (α1-AT deficiency) with a PiZZ genotype are at high risk to develop fatal emphysema during adulthood. Spirometry is commonly used for monitoring these patients. However early manifestation of emphysema is suspected within the peripheral airways that are not accessible by spirometry. We hypothesized that the Lung Clearance Index (LCI) from Multiple Breath Nitrogen Washout (MBW_{N2}) tests is sensitive for detecting ventilation inhomogeneity (VI) within the peripheral airways and may thus be suitable to bridge this diagnostic gap. We present preliminary data of an ongoing multi centre study assessing the clinical value of measuring the LCI in patients with α1-AT deficiency. 89 patients (4-73 years, PiZZ) and 40 controls (7-84years) performed 2-3 single MBW_{N2} (EasyOne Pro LAB™) and subsequent spirometry. Mean (SD) LCI in patients was 9.0 (2.9) and 6.5 (0.72) in controls. Mean difference (95% ci, p-value) between the groups was -2.49 (-3.14;-1.84, <0.001). Mean FEV_1-z (SD) in patients was -1.36 (1.8). In 31/89 patients LCI and FEV_1-z were abnormal, in 32/89 LCI and FEV_1-z were normal, in 1/89 FEV_1-z was abnormal while LCI was normal and in 25/89 LCI was abnormal while FEV_1-z remained normal. LCI discriminated significantly between patients and controls. 44% of the patients with a normal FEV_1-z had an abnormal LCI potentially indicating early manifestation of α1-AT deficiency related lung disease that is not detectable using spirometry alone. We conclude from these

preliminary data that the LCI reflects presence and extent of α1-AT deficiency related lung disease and detects these changes earlier than spirometry. This project is supported by Grifols.