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The assessment of short- and long-term changes in lung function in cystic fibrosis using ^{129}Xe MRI

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^{129}Xe-MRI in CF is highly repeatable. In patients with normal FEV_1, ^{129}Xe-MRI is also sensitive to detect changes in longitudinal lung function and should be highly informative in an era of CFTR modulators and increasingly preserved FEV_1 https://bit.ly/2C0D8Np

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ABSTRACT

Introduction: Xenon-129 (^{129}Xe) ventilation magnetic resonance imaging (MRI) is sensitive to detect early cystic fibrosis (CF) lung disease and response to treatment. ^{129}Xe-MRI could play a significant role in clinical trials and patient management. Here we present data on the repeatability of imaging measurements and their sensitivity to longitudinal change.

Methods: 29 children and adults with CF and a range of disease severity were assessed twice, a median (interquartile range (IQR)) of 16.0 (14.4–19.5) months apart. Patients underwent ^{129}Xe-MRI, lung clearance index (LCI), body plethysmography and spirometry at both visits. 11 patients repeated ^{129}Xe-MRI in the same session to assess the within-visit repeatability. The ventilation defect percentage (VDP) was the primary metric calculated from ^{129}Xe-MRI.

Results: At baseline, mean±SD age was 23.0±11.1 years and forced expiratory volume in 1 s (FEV_1) z-score was −2.2±2.0. Median (IQR) VDP was 9.5 (3.4–31.6)% and LCI was 9.0 (7.7–13.7). Within- and inter-visit repeatability of VDP was high. At 16 months there was no single trend of ^{129}Xe-MRI disease progression. Visible ^{129}Xe-MRI ventilation changes were common, which reflected changes in VDP. Based on the within-visit repeatability, a significant short-term change in VDP is >±1.6%. For longer-term follow-up, changes in VDP of up to ±7.7% can be expected, or ±4.1% for patients with normal FEV_1. No patient had a significant change in FEV_1; however, 59% had change in VDP >±1.6%. In patients with normal FEV_1, there were significant changes in ventilation and in VDP.

Conclusions: ^{129}Xe-MRI is a highly effective method for assessing longitudinal lung disease in patients with CF. VDP has great potential as a sensitive clinical outcome measure of lung function and end-point for clinical trials.