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Magnetic Resonance Imaging of the Lung for Follow-up and Therapy Control in Patients with Cystic Fibrosis

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Recent studies demonstrated that structural lung disease in infants and young children with CF begins early, often even in the absence of clinical symptoms. For a long time chest X-ray and/or CT imaging considered the “gold standard” for CF patients. However, the use of CT for lifelong follow-up should be restricted due to the high cumulative radiation dose. Thus non-invasive detection and monitoring of early CF lung disease remains challenging. In addition, CT usually does not provide the required sensitivity for differentiating between acute and chronic changes in the CF lung, especially in the early stages of the disease.

Recent developments in the field of MRI, enables CT-like, morphological imaging of the lungs. In addition, MRI can offer better differentiation of soft tissue and is able to represent the functional information of the lung parenchyma primarily with regard to its perfusion.

Several previous studies compared MRI and CT in adult CF patients and showed MRI to be sensitive enough for CF lung imaging yielding equivalent diagnostic results. The current work focuses on infants and young children with early CF lung disease. It was hypothesized that MRI would be able to visualize the morphological and functional lung changes at the early stages of CF lung disease in young patients. Therefore, it should be possible to demonstrate that MRI also allows for therapy monitoring in cases of exacerbation in infants and young children. For this reason, the results of MRI examinations in children with and without CF were compared. In addition, MRI examinations before and after antibiotic therapy in children with exacerbation of CF lung disease were compared.

A total of 40 infants and young children with stable CF lung disease and 10 infants and young children with exacerbation of CF lung disease were enrolled in this study. For comparison, 10 infants and young children patients without lung disease were examined using MRI.

For morphological imaging T1- and T2-weighted sequences were used. T1-weighted TSE sequence in free breathing was used before and after the intravenous administration of contrast media. The T2-weighted sequence was used was a HASTE sequence with ECG gating in an inspiratory breath hold or with navigator triggering in free breathing depending on the degree of cooperation of the patient. Lung perfusion images were acquired with a time-resolved 3D gradient echo pulse sequence (Fast low angle shot, FLASH) during the intravenous administration of the contrast agent.

In order to allow for a better comparison of MRI findings, the evaluation was based on a morpho-functional MRI score. Therefore, lobe-based analysis was performed for the following parameters: bronchiectasis/wall thickening, mucus plugging, abscesses/sacculations, consolidations, special findings and perfusion defects.

MRI demonstrated bronchial wall thickening/bronchiectasis, mucus plugging and perfusion deficits from the first year of life in most stable patients with CF (global score 10.0 ± 4.0). No morphological or functional changes in the lung of non-CF controls were found (score 0.0 ± 0.0 ; $P < 0.001$). In CF patients with exacerbations, the global MRI score was increased to 18.0 ± 2.0 ($P < 0.001$), and was significantly reduced to 12.0 ± 3.0 ($P < 0.05$) after antibiotic therapy.

In conclusion, MRI is capable of visualizing subtle morphologic and functional lung changes already in the very early stage of CF lung disease. In contrast to chest X-ray or CT, MRI allows for the differentiation between active and chronic pulmonary changes as well as monitoring the changes before and after therapy. Therefore, in contrast to the chest X-ray or CT, MRI is arguably a more suitable method for the visualization of morphological and functional lung changes throughout the disease and for monitoring the effects of different therapies in newborn and young children with CF.