

The Clinical Use of Lung MRI in Cystic Fibrosis

What, Now, How?

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e-Appendix 1

The structural alterations in CF that can be objectively identified using imaging are the following¹:

- bronchiectasis is an irreversible dilatation of the airway lumen; wall thickening is an enlargement of the bronchial wall thickness, secondary to either acute inflammation or chronic remodeling

- mucus plug is a secretion of viscosity secondary to an inflammatory exudate or altered airway clearance or both

- atelectasis is the reduction in inflation of all or part of the lung, secondary to bronchial obstruction

- consolidation refers to exudate or other disease product that replace alveolar air; emphysema is the destruction of the lung parenchyma

- bulla is an isolated pocket of air within the parenchyma.

Some structural abnormalities do not have an official definition, such as sacculation or infiltrate. The main hallmarks in CF are, however, bronchiectasis, wall thickening, mucus plugs, and consolidations.

e-Appendix 2

For practical implementation, the lung MRI procedure may be as follows:

- It is highly beneficial to meet the patients before their MR scan, especially those for whom it is the first time. The main oral information to be provided are the rationale to use MRI, the longer acquisition time than CXR or CT, a noisy procedure, the necessity to avoid other body movements than the respiratory movements. For children, a training session outside the MR scan, using MR-like toys, are beneficials. Ideally this should be done by lung function technicians. The same person that gives the breathing instruction should also give the breathing instructions during the MRI session. Especially in children this reduces anxiety, reduces movement artefacts, and especially improves the quality of breath hold maneuvers at residual volume². The latter maximizes the contrast between low intensity regions an normal hyper intensity regions.
- A music style choice can be discussed as an entertainment during the MR procedure, although a relaxing music style is preferred to avoid unwanted body movements, especially in kids.
- Once the patient is inside the MR scan, the first step is to acquire a survey, in a couple of seconds.
 For those patients who would perform lung MRI for the first time, it can be beneficial to spend some times in starting a MR sequence or two as training, that enable the patient to better figure out what

the MR procedure consists in. Sometimes, a patient may unexpectedly discover that he/she suffers Online supplements are not copyedited prior to posting and the author(s) take full responsibility for the accuracy of all data.

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from claustrophobia inside the MR scan and thus, the examination has to be stopped as a contraindication.

- The second step is to implement the MR sequences themselves, in terms of technical parameters (field of view, matrix, slice thickness, TR TEs, etc). Standard pre registered MR protocols are advised. Assistance by a MR engineer is useful for first attempts.
- The order of sequences does not matter. However, it can be said that all MR sequences that are acquired in free-breathing with or without a respiratory synchronization system (UTE, T2w, Fourier) can be chained one after another without any further intervention by the radiographer after starting the process. In our proposal for lung MR protocol, only the T1w sequence requires a 15 seconds breath-hold maneuver, and can be placed either at the beginning or at the end of the core protocol.
- It is advised to supervise the results step by step and sometimes repeat the acquisition procedure, in case of artifacts.
- In our institution, the acceptance of the imaging modality by the CF patients is high. In a survey completed by 30 adults with CF, 29/30 answered "yes" to the question: "would you go on with MRI only for the next yearly imaging evaluations?" (unpublished data). In children, things are sometimes more progressive. We use it as a routine procedure at the age of 6-year-old. Some kids may feel afraid of MRI at first so that we complete both MRI and CT for the first attempt. However, our experience show that the same kids will progressively accept it much more during the next procedures, as an already completed examination, enabling to progressively switch to this radiation-free procedure. In infants and preschool children, it has been reported in the literature that either CT or MRI may require sedation.
- Finally, an understanding of the basics of the MR procedure and the rationale of its reading is required. Indeed, acceptance by the clinician often comes along with its understanding. This literature review has been written by keeping this in mind. The participation of radiologists to multidisciplinary meetings may also be useful.

Supplemental References

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