

Evidence Discussion (CH 7.2)

Adult Cystic Fibrosis Evidence Summary

Imaging is an important method of evaluating the lungs in people with cystic fibrosis (CF). It has a stronger correlation with disease severity than pulmonary function tests and facilitates prompt therapy which may help limit irreversible lung damage.(1) Chest x- ray (CR) is less sensitive than CT chest at detecting early structural changes and disease progression.(2) However, CR is still most commonly used as the first line imaging examination for the assessment of acute complications due to its low cost, availability, low radiation and speed of acquisition.(3) CT is increasingly being used to monitor disease progression and make treatment decisions, but the routine use of CT for short term follow up during pulmonary exacerbations is not recommended due to the risk of a high cumulative radiation dose. Low dose chest CT (LDCT) is useful in patients with persistent respiratory symptoms and decreased lung function despite appropriate therapy.(2) There is little evidence regarding the optimal timing of CT monitoring. The current best clinical practice in several European CF centers is a CT every two years with a radiation dose as low as reasonably achievable (ALARA). Follow up imaging is determined by individual patient-dependent clinical factors.(2) The CF Foundation guidelines for adult CF clinical care recommend CR every 2-4 years in those with a stable clinical status and state that imaging should be considered if there are symptoms or signs of an acute pulmonary exacerbation, pneumothorax, lobar atelectasis or hemoptysis.(4,5)

Several emerging techniques offer promising means of pulmonary imaging using less ionizing radiation, including ultra-low dose CT (ULDCT) and MRI. (1,6) The radiation dose with CR is 0.02mSv, 5.4 mSv for standard dose CT, 1 to 2 mSv for LDCT, and 0.05-0.08mSv for ULDCT. While pulmonary MRI has promise as a means of routinely monitoring CF lung disease, it is currently limited by a lack of availability, high cost, lack of validation and standardized protocols, and the need for sedation or anesthesia in some patients.(1,2)

References

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