ONLINE SUPPLEMENT

Appendix 1

Research Strategy

General

(Cystic fibrosis OR pulmonary cystic fibrosis) AND imaging AND (Randomized controlled trial OR controlled clinical trials, non-randomized OR clinical trials, non-randomized OR Systematic review OR review, systematic OR meta-analysis OR Network meta-analysis OR Guideline OR Clinical practice guideline OR Consensus OR Protocol)

(Generates 153 hits)

(Cystic fibrosis OR pulmonary cystic fibrosis) AND imaging AND (pulmonary disease OR lung disease) AND (Randomized controlled trial OR controlled clinical trials, non-randomized OR clinical trials, non-randomized OR Systematic review OR review, systematic OR meta-analysis OR Network meta-analysis OR Guideline OR Clinical practice guideline OR Consensus OR Protocol)

(Generates 119 hits)

(Cystic fibrosis OR pulmonary cystic fibrosis) AND imaging AND (pulmonary disease OR lung disease) AND (Patient compliance OR patient cooperation)

(Generates 1 hit)

Lung ultrasound

(Cystic fibrosis OR pulmonary cystic fibrosis) AND (imaging OR radiology OR radiology department, hospital OR Radiologists OR radiography) AND (Ultrasonography OR Diagnostic ultrasound OR Ultrasound imaging OR Ultrasonography, Doppler OR) AND (Disease progression OR Disease exacerbation OR Acute lung injury OR Respiratory function tests OR Interstitial syndrome OR Pleural effusion OR Emphysema OR Pulmonary emphysema) AND (Randomized controlled trial OR controlled clinical trials, non-randomized OR clinical trials, non-randomized OR systematic review OR review, systematic OR meta-analysis OR Network meta-analysis OR Guideline OR Clinical practice guideline OR Consensus OR Protocol)

(Generates 49 hits)

Chest Radiographs

(Cystic fibrosis OR pulmonary cystic fibrosis) AND (imaging OR radiology OR radiology department, hospital OR Radiologists OR radiography) AND (Chest X-ray OR Chest radiography) AND (Randomized controlled trial OR controlled clinical trials, non-randomized OR clinical trials, non-randomized OR systematic review OR review, systematic OR meta-analysis OR Network meta-analysis OR Guideline OR Clinical practice guideline OR Consensus OR Protocol)

(Generates 65 hits)

(Cystic fibrosis OR pulmonary cystic fibrosis) AND (imaging OR radiology OR radiology department, hospital OR Radiologists OR radiography) AND (Chest X-ray OR Chest radiography OR Radiation dosage OR Dose fractionation, radiation) AND (Disease progression OR Disease exacerbation OR Acute lung injury OR Respiratory function tests) AND (Randomized controlled trial OR controlled clinical trials, non-randomized OR clinical trials, non-randomized OR systematic review OR review, systematic OR meta-analysis OR Network meta-analysis OR Guideline OR Clinical practice guideline OR Consensus OR Protocol)

(Generates 38 hits)

Computed tomography

(Cystic fibrosis OR pulmonary cystic fibrosis) AND (imaging OR radiology OR radiology department, hospital OR Radiologists OR radiography) AND (Tomography, X-ray computed OR Tomography, spiral computed OR Cone-Beam Computed Tomography OR Positron emission tomography computed tomography OR Tomography, emission-computed, single-photon OR Radiation dosage OR Dose fractionation, radiation) AND (Disease progression OR Disease exacerbation OR Acute lung injury OR Respiratory function tests) AND (Randomized controlled trial OR controlled clinical trials, non-randomized OR clinical trials, non-randomized OR systematic review OR review, systematic OR meta-analysis OR Network meta-analysis OR Guideline OR Clinical practice guideline OR Consensus OR Protocol)

(Generates 34 hits)

(Cystic fibrosis OR pulmonary cystic fibrosis) AND (imaging OR radiology OR radiology department, hospital OR Radiologists OR radiography) AND (Tomography, X-ray computed OR Tomography, spiral computed OR Cone-Beam Computed Tomography OR Positron emission tomography computed tomography OR Tomography, emission-computed, single-photon OR Radiation dosage OR Dose fractionation, radiation) AND (Tomography scanners, X-ray computed OR Multidetector computed tomography OR Image processing, computer-assisted OR Image acquisition OR Image interpretation, computer-assisted OR Radiographic image interpretation, computer-assisted OR Radiographic image enhancement) AND (Randomized controlled trial OR controlled clinical trials, non-randomized OR clinical trials, non-randomized OR systematic review OR review, systematic OR meta-analysis OR Network meta-analysis OR Guideline OR Clinical practice guideline OR Consensus OR Protocol)

(Generates 68 hits)

Magnetic resonance imaging

(Cystic fibrosis OR pulmonary cystic fibrosis) AND (imaging OR radiology OR radiology department, hospital OR Radiologists OR radiography) AND (Magnetic resonance imaging OR Diffusion magnetic resonance imaging OR Functional magnetic resonance imaging OR Contrast media OR Image processing, computer-assisted OR Image acquisition OR Image interpretation, computer-assisted) AND (Disease progression OR Disease exacerbation OR Acute lung injury OR Respiratory function tests) AND (Randomized controlled trial OR controlled clinical trials, non-randomized OR clinical trials, non-randomized OR systematic review OR review, systematic OR meta-analysis OR Network meta-analysis OR Guideline OR Clinical practice guideline OR Consensus OR Protocol)

(Generates 41 hits)

Cooperation between radiologists and clinicians

(Cystic fibrosis OR pulmonary cystic fibrosis) AND (imaging OR radiology OR radiology department, hospital OR Radiologists) AND (Multidisciplinary OR Interdisciplinary communication)

(Generates 26 hits)

(Cystic fibrosis OR pulmonary cystic fibrosis) AND (imaging OR radiology OR radiology department, hospital OR Radiologists) AND (Multidisciplinary OR Interdisciplinary communication) AND

(Randomized controlled trial OR controlled clinical trials, non-randomized OR clinical trials, non-randomized OR systematic review OR review, systematic OR meta-analysis OR Network meta-analysis OR Guideline OR Clinical practice guideline OR Consensus OR Protocol)

(Generates 8 hits)

Online Supplement

Title: Lung imaging in Cystic Fibrosis: consensus statement and recommendations for pulmonologists and radiologists from the "iMAging managEment of cySTic fibROsis" (MAESTRO) consortium

1E List of selected articles

1. Amaxopoulou C, Gnannt R, Higashigaito K, et al (2018) Structural and perfusion magnetic resonance imaging of the lung in cystic fibrosis. Pediatr Radiol 48:165–175. https://doi.org/10.1007/s00247-017-4021-8
2. Bayfield K, Weinheimer O, Boyton C, et al (2020) Late Breaking Abstract - Use of ultra-low dose CT does not impact structure-function relationships in early Cystic Fibrosis lung disease. In: Cystic fibrosis. European Respiratory Society, p 4310
3. Bayfield KJ, Douglas TA, Rosenow T, et al (2021) Time to get serious about the detection and monitoring of early lung disease in cystic fibrosis. Thorax thoraxjnl-2020-216085. https://doi.org/10.1136/thoraxjnl-2020-216085
4. Behrendt L, Voskrebenzev A, Klimeš F, et al (2020) Validation of Automated Perfusion-Weighted Phase-Resolved Functional Lung (PREFUL)-MRI in Patients With Pulmonary Diseases. J Magn Reson Imaging 52:103–114. https://doi.org/10.1002/jmri.27027
5. Belessis Y, Dixon B, Hawkins G, et al (2012) Early cystic fibrosis lung disease detected by bronchoalveolar lavage and lung clearance index. Am J Respir Crit Care Med 185:862–873. https://doi.org/10.1164/rccm.201109-1631OC
6. Bell SC, Mall MA, Gutierrez H, et al (2020) The future of cystic fibrosis care: a global perspective. Lancet Respir Med 8:65–124. https://doi.org/10.1016/S2213-2600(19)30337-6
7. Benden C (2005) The Chrispin-Norman score in cystic fibrosis: doing away with the lateral view. Eur Respir J 26:894–897. https://doi.org/10.1183/09031936.05.00059105
8. Benlala I, Hocke F, Macey J, et al (2020) Quantification of MRI T2-weighted High Signal Volume in Cystic Fibrosis: A Pilot Study. Radiology 294:186–196. https://doi.org/10.1148/radiol.2019190797

9. Benlala I, Point S, Leung C, et al (2020) Volumetric quantification of lung MR signal intensities using ultrashort TE as an automated score in cystic fibrosis. Eur Radiol 30:5479–5488. https://doi.org/10.1007/s00330-020-06910-w
10. Bhalla M, Turcios N, Aponte V, et al (1991) Cystic fibrosis: scoring system with thin-section CT. Radiology 179:783–788. https://doi.org/10.1148/radiology.179.3.2027992
11. Bortoluzzi C-F, Volpi S, D'Orazio C, et al (2014) Bronchiectases at early chest computed tomography in children with cystic fibrosis are associated with increased risk of subsequent pulmonary exacerbations and chronic pseudomonas infection. J Cyst Fibros 13:564–571. https://doi.org/10.1016/j.jcf.2014.03.006
12. Bortoluzzi CF, Pontello E, Pintani E, et al (2020) The impact of chest computed tomography and chest radiography on clinical management of cystic fibrosis lung disease. J Cyst Fibros 19:641–646. https://doi.org/10.1016/j.jcf.2019.08.005
13. Bouma NR, Janssens HM, Andrinopoulou E, Tiddens HAWM (2020) Airway disease on chest computed tomography of preschool children with cystic fibrosis is associated with school-age bronchiectasis. Pediatr Pulmonol 55:141–148. https://doi.org/10.1002/ppul.24498
14. Brenner DJ, Hall EJ (2007) Computed Tomography — An Increasing Source of Radiation Exposure. N Engl J Med 357:2277–2284. https://doi.org/10.1056/NEJMra072149
15. Brody AS (2004) Scoring Systems for CT in Cystic Fibrosis: Who Cares? Radiology 231:296–298. https://doi.org/10.1148/radiol.2312032097
16. Brody AS, Klein JS, Molina PL, et al (2004) High-resolution computed tomography in young patients with cystic fibrosis: Distribution of abnormalities and correlation with pulmonary function tests. J Pediatr 145:32–38. https://doi.org/10.1016/j.jpeds.2004.02.038
17. Brody AS, Kosorok MR, Li Z, et al (2006) Reproducibility of a Scoring System for Computed Tomography Scanning in Cystic Fibrosis. J Thorac Imaging 21:14–21. https://doi.org/10.1097/01.rti.0000203937.82276.ce
18. Brody AS, Molina PL, Klein JS, et al (1999) High-resolution computed tomography of the chest in children with cystic fibrosis: support for use as an outcome surrogate. Pediatr Radiol 29:731–735. https://doi.org/10.1007/s002470050684
19. Brody AS, Sucharew H, Campbell JD, et al (2005) Computed Tomography Correlates with Pulmonary Exacerbations in Children with Cystic Fibrosis. Am J Respir Crit Care Med 172:1128–1132. https://doi.org/10.1164/rccm.200407-989OC

20. Byrnes CA, Vidmar S, Cheney JL, et al (2013) Prospective evaluation of respiratory exacerbations in children with cystic fibrosis from newborn screening to 5 years of age. Thorax 68:643–651. https://doi.org/10.1136/thoraxjnl-2012-202342
21. Cademartiri F, Luccichenti G, Palumbo AA, et al (2008) Predictive Value of Chest CT in Patients with Cystic Fibrosis: A Single-Center 10-Year Experience. Am J Roentgenol 190:1475–1480. https://doi.org/10.2214/AJR.07.3000
22. Caley L, Smith L, White H, Peckham DG (2021) Average rate of lung function decline in adults with cystic fibrosis in the United Kingdom: Data from the UK CF registry. J Cyst Fibros 20:86–90. https://doi.org/10.1016/j.jcf.2020.04.008
23. Chassagnon G, Martin C, Burgel P-R, et al (2018) An automated computed tomography score for the cystic fibrosis lung. Eur Radiol 28:5111–5120. https://doi.org/10.1007/s00330-018-5516-x
24. Ciet P, Bertolo S, Ros M, et al (2017) Detection and monitoring of lung inflammation in cystic fibrosis during respiratory tract exacerbation using diffusion-weighted magnetic resonance imaging. Eur Respir J 50:1601437. https://doi.org/10.1183/13993003.01437-2016
25. Ciet P, Serra G, Andrinopoulou ER, et al (2016) Diffusion weighted imaging in cystic fibrosis disease: beyond morphological imaging. Eur Radiol 26:3830–3839. https://doi.org/10.1007/s00330-016-4248-z
26. Ciet P, Serra G, Bertolo S, et al (2016) Assessment of CF lung disease using motion corrected PROPELLER MRI: a comparison with CT. Eur Radiol 26:780–787. https://doi.org/10.1007/s00330-015-3850-9
27. Ciet P, Tiddens HAWM, Wielopolski PA, et al (2015) Magnetic resonance imaging in children: common problems and possible solutions for lung and airways imaging. Pediatr Radiol. https://doi.org/10.1007/s00247-015-3420-y
28. Cohen-Cymberknoh M, Shoseyov D, Kerem E (2011) Managing cystic fibrosis: strategies that increase life expectancy and improve quality of life. Am J Respir Crit Care Med 183:1463–71. https://doi.org/10.1164/rccm.201009-1478CI
29. Cohen-Cymberknoh M, Ben Meir E, Gartner S, et al (2021) How abnormal is the normal? Clinical characteristics of CF patients with normal FEV 1. Pediatr Pulmonol ppul.25371. https://doi.org/10.1002/ppul.25371

30. Conway S, Balfour-Lynn IM, De Rijcke K, et al (2014) European Cystic Fibrosis Society Standards of Care: Framework for the Cystic Fibrosis Centre. J Cyst Fibros 13:S3–S22. https://doi.org/10.1016/j.jcf.2014.03.009
31. Couch MJ, Ball IK, Li T, et al. (2019) 19 F MRI of the Lungs Using Inert Fluorinated Gases: Challenges and New Developments. J Magn Reson Imaging 49:343–354. doi: 10.1002/jmri.26292
32. Crowley C, Connor OJO, Ciet P, et al (2021) The evolving role of radiological imaging in cystic fibrosis. Curr Opin Pulm Med 27:575–585. https://doi.org/10.1097/MCP.0000000000000828
33. Davies G, Thia LP, Stocks J, et al (2020) Minimal change in structural, functional and inflammatory markers of lung disease in newborn screened infants with cystic fibrosis at one year. J Cyst Fibros 19:896–901. https://doi.org/10.1016/j.jcf.2020.01.006
34. Davis SD, Fordham LA, Brody AS, et al (2007) Computed Tomography Reflects Lower Airway Inflammation and Tracks Changes in Early Cystic Fibrosis. Am J Respir Crit Care Med 175:943–950. https://doi.org/10.1164/rccm.200603-343OC
35. de Jong PA, Achterberg J A, Kessels O a M, et al (2011) Modified Chrispin-Norman chest radiography score for cystic fibrosis: observer agreement and correlation with lung function. Eur Radiol 21:722–9. https://doi.org/10.1007/s00330-010-1972-7
36. de Jong PA, Lindblad A, Rubin L, et al (2006) Progression of lung disease on computed tomography and pulmonary function tests in children and adults with cystic fibrosis. Thorax 61:80–5. https://doi.org/10.1136/thx.2005.045146
37. de Jong PA, Mayo JR, Golmohammadi K, et al (2006) Estimation of Cancer Mortality Associated with Repetitive Computed Tomography Scanning. Am J Respir Crit Care Med 173:199–203. https://doi.org/10.1164/rccm.200505-810OC
38. de Jong PA, Nakano Y, Lequin MH, et al (2004) Progressive damage on high resolution computed tomography despite stable lung function in cystic fibrosis. Eur Respir J 23:93–97. https://doi.org/10.1183/09031936.03.00006603
39. Dournes G, Berger P, Refait J, et al (2017) Allergic Bronchopulmonary Aspergillosis in Cystic Fibrosis: MR Imaging of Airway Mucus Contrasts as a Tool for Diagnosis. Radiology 285:261–269. https://doi.org/10.1148/radiol.2017162350

40. Dournes G, Hall CS, Willmering MM, et al (2021) Artificial intelligence in CT for quantifying lung changes in the era of CFTR modulators. Eur Respir J 2100844. https://doi.org/10.1183/13993003.00844-2021
41. Dournes G, Menut F, Macey J, et al (2016) Lung morphology assessment of cystic fibrosis using MRI with ultra-short echo time at submillimeter spatial resolution. Eur Radiol. https://doi.org/10.1007/s00330-016-4218-5
42. Eichinger M, Optazaite D-E, Kopp-Schneider A, et al (2012) Morphologic and functional scoring of cystic fibrosis lung disease using MRI. Eur J Radiol 81:1321–9. https://doi.org/10.1016/j.ejrad.2011.02.045
43. Eichinger M, Tetzlaff R, Puderbach M, et al (2007) Proton magnetic resonance imaging for assessment of lung function and respiratory dynamics. Eur J Radiol 64:329–34. https://doi.org/10.1016/j.ejrad.2007.08.007
44. Ellemunter H, Fuchs SI, Unsinn KM, et al (2010) Sensitivity of lung clearance index and chest computed tomography in early cf lung disease. Respir Med 104:1834–1842. https://doi.org/10.1016/j.rmed.2010.06.010
45. Ernst CW, Basten IA, Ilsen B, et al (2014) Pulmonary Disease in Cystic Fibrosis: Assessment with Chest CT at Chest Radiography Dose Levels. Radiology 273:597–605. https://doi.org/10.1148/radiol.14132201
46. Fleischer S, Kraus MS, Gatidis S, et al (2020) New severity assessment in cystic fibrosis: signal intensity and lung volume compared to LCI and FEV1: preliminary results. Eur Radiol 30:1350–1358. https://doi.org/10.1007/s00330-019-06462-8
47. Fuchs SI, Gappa M, Eder J, et al (2014) Tracking Lung Clearance Index and chest CT in mild cystic fibrosis lung disease over a period of three years. Respir Med 108:865–874. https://doi.org/10.1016/j.rmed.2014.03.011
48. Garg MK, Gupta P, Agarwal R, et al (2015) MRI: a new paradigm in imaging evaluation of allergic bronchopulmonary aspergillosis? Chest 147:e58–e59. https://doi.org/10.1378/chest.14-2347
49. Gilchrist FJ, Buka R, Jones M, et al (2018) Clinical indications and scanning protocols for chest CT in children with cystic fibrosis: a survey of UK tertiary centres. BMJ Paediatr Open 2:e000367. https://doi.org/10.1136/bmjpo-2018-000367

50. Glandorf J, Klimeš F, Behrendt L, et al (2021) Perfusion quantification using voxel-wise proton density and median signal decay in PREFUL MRI. Magn Reson Med mrm.28787. https://doi.org/10.1002/mrm.28787
51. Goralski JL, Chung SH, Glass TM, et al. (2020) Dynamic perfluorinated gas MRI reveals abnormal ventilation despite normal FEV1 in cystic fibrosis. JCI Insight. doi: 10.1172/jci.insight.133400
52. Goralski JL, Stewart NJ, Woods JC (2021) Novel imaging techniques for cystic fibrosis lung disease. Pediatr Pulmonol 56 Suppl 1:S40–S54. https://doi.org/10.1002/ppul.24931
53. Goris ML, Zhu HJ, Blankenberg F, et al (2003) An Automated Approach to Quantitative Air Trapping Measurements in Mild Cystic Fibrosis. Chest 123:1655–1663. https://doi.org/10.1378/chest.123.5.1655
54. Graeber SY, Boutin S, Wielpütz MO, et al (2021) Effects of Lumacaftor–Ivacaftor on Lung Clearance Index, Magnetic Resonance Imaging, and Airway Microbiome in Phe508del Homozygous Patients with Cystic Fibrosis. Ann Am Thorac Soc 18:971–980. https://doi.org/10.1513/AnnalsATS.202008-1054OC
55. Grasemann H, Ciet P, Amin R, et al (2017) Changes in magnetic resonance imaging scores and ventilation inhomogeneity in children with cystic fibrosis pulmonary exacerbations. Eur Respir J 50:1700244. https://doi.org/10.1183/13993003.00244-2017
56. Greene KE, Takasugi JE, Godwin JD, et al (1994) Radiographic changes in acute exacerbations of cystic fibrosis in adults: a pilot study. AJR Am J Roentgenol 163:557–62. https://doi.org/10.2214/ajr.163.3.8079843
57. Gustafsson PM, De Jong PA, Tiddens HAWM, Lindblad A (2007) Multiple-breath inert gas washout and spirometry versus structural lung disease in cystic fibrosis. Thorax 63:129–134. https://doi.org/10.1136/thx.2007.077784
58. Hall GL, Logie KM, Parsons F, et al (2011) Air Trapping on Chest CT Is Associated with Worse Ventilation Distribution in Infants with Cystic Fibrosis Diagnosed following Newborn Screening. PLoS One 6:e23932. https://doi.org/10.1371/journal.pone.0023932
59. Hirsch FW, Sorge I, Vogel-Claussen J, et al (2020) The current status and further prospects for lung magnetic resonance imaging in pediatric radiology. Pediatr Radiol 50:734–749. https://doi.org/10.1007/s00247-019-04594-z

60. Horsley AR, Davies JC, Gray RD, et al (2013) Changes in physiological, functional and structural markers of cystic fibrosis lung disease with treatment of a pulmonary exacerbation. Thorax 68:532–539. https://doi.org/10.1136/thoraxjnl-2012-202538
61. Hota P, Madan R (2020) Cystic Fibrosis from Childhood to Adulthood. Radiol Clin North Am 58:475–486. https://doi.org/10.1016/j.rcl.2019.12.003
62. Huda W (2007) Radiation Doses and Risks in Chest Computed Tomography Examinations. Proc Am Thorac Soc 4:316–320. https://doi.org/10.1513/pats.200611-172HT
63. Judge EP, Dodd JD, Masterson JB, Gallagher CG (2006) Pulmonary Abnormalities on High-Resolution CT Demonstrate More Rapid Decline Than FEV 1 in Adults With Cystic Fibrosis. Chest 130:1424–1432. https://doi.org/10.1378/chest.130.5.1424
64. Kerem E, Reisman J, Corey M, et al (1992) Prediction of Mortality in Patients with Cystic Fibrosis. N Engl J Med 326:1187–1191. https://doi.org/10.1056/NEJM199204303261804
65. Kino A, Zucker EJ, Honkanen A, et al (2019) Ultrafast pediatric chest computed tomography: comparison of free-breathing vs. breath-hold imaging with and without anesthesia in young children. Pediatr Radiol 49:301–307. https://doi.org/10.1007/s00247-018-4295-5
66. Konietzke P, Weinheimer O, Wielpütz MO, et al (2018) Validation of automated lobe segmentation on paired inspiratory-expiratory chest CT in 8-14 year-old children with cystic fibrosis. PLoS One 13:e0194557. https://doi.org/10.1371/journal.pone.0194557
67. Kuo W, Andrinopoulou E-R, Perez-Rovira A, et al (2016) Objective airway artery dimensions compared to CT scoring methods assessing structural cystic fibrosis lung disease. J Cyst Fibros. https://doi.org/10.1016/j.jcf.2016.05.015
68. Kuo W, Ciet P, Tiddens H a WM, et al (2014) Monitoring Cystic Fibrosis Lung Disease by Computed Tomography. Radiation Risk in Perspective. Am J Respir Crit Care Med 189:1328–1336. https://doi.org/10.1164/rccm.201311-2099CI
69. Kuo W, de Bruijne M, Petersen J, et al (2017) Diagnosis of bronchiectasis and airway wall thickening in children with cystic fibrosis: Objective airway-artery quantification. Eur Radiol 27:4680–4689. https://doi.org/10.1007/s00330-017-4819-7
70. Kuo W, Kemner-van de Corput MPC, Perez-Rovira A, et al (2016) Multicentre chest computed tomography standardisation in children and adolescents with cystic fibrosis: the way forward. Eur Respir J 47:1706–1717. https://doi.org/10.1183/13993003.01601-2015

71. Leutz-Schmidt P, Eichinger M, Stahl M, et al (2019) Ten years of chest MRI for patients with cystic fibrosis. Radiologe 59:10–20. https://doi.org/10.1007/s00117-019-0553-2
72. Leutz-Schmidt P, Stahl M, Sommerburg O, et al (2018) Non-contrast enhanced magnetic resonance imaging detects mosaic signal intensity in early cystic fibrosis lung disease. Eur J Radiol 101:178–183. https://doi.org/10.1016/j.ejrad.2018.02.023
73. Lin S, Lin M, Lau KK (2019) Image quality comparison between model-based iterative reconstruction and adaptive statistical iterative reconstruction chest computed tomography in cystic fibrosis patients. J Med Imaging Radiat Oncol 63:602–609. https://doi.org/10.1111/1754-9485.12895
74. Loeve M, Gerbrands K, Hop WC, et al (2011) Bronchiectasis and pulmonary exacerbations in children and young adults with cystic fibrosis. Chest 140:178–85. https://doi.org/10.1378/chest.10-1152
75. Loeve M, Lequin MH, de Bruijne M, et al (2009) Cystic Fibrosis: Are Volumetric Ultra-Low-Dose Expiratory CT Scans Sufficient for Monitoring Related Lung Disease? Radiology 253:223–229. https://doi.org/10.1148/radiol.2532090306
76. Loeve M, Rosenow T, Gorbunova V, et al (2015) Reversibility of trapped air on chest computed tomography in cystic fibrosis patients. Eur J Radiol 84:1184–1190. https://doi.org/10.1016/j.ejrad.2015.02.011
77. Loeve M, van Hal PTW, Robinson P, et al (2009) The spectrum of structural abnormalities on CT scans from patients with CF with severe advanced lung disease. Thorax 64:876–82. https://doi.org/10.1136/thx.2008.110908
78. Malviya S, Voepel-Lewis T, Eldevik OP, et al (2000) Sedation and general anaesthesia in children undergoing MRI and CT: adverse events and outcomes. Br J Anaesth 84:743–748. https://doi.org/10.1093/oxfordjournals.bja.a013586
79. Marshall H, Horsley A, Taylor CJ, et al (2017) Detection of early subclinical lung disease in children with cystic fibrosis by lung ventilation imaging with hyperpolarised gas MRI. Thorax 72:760–762. https://doi.org/10.1136/thoraxjnl-2016-208948
80. Martini K, Gygax CM, Benden C, et al (2018) Volumetric dynamic oxygen-enhanced MRI (OE-MRI): comparison with CT Brody score and lung function in cystic fibrosis patients. Eur Radiol 28:4037–4047. https://doi.org/10.1007/s00330-018-5383-5

81. Min-Zhao Lee, Weidong Cai, Yang Song, et al (2013) Fully automated scoring of chest radiographs in cystic fibrosis. In: 2013 35th Annual International Conference of the IEEE Engineering in Medicine and Biology Society (EMBC). IEEE, pp 3965–3968
82. Moloney F, Kavanagh RG, Ronan NJ, et al (2021) Ultra-low-dose thoracic CT with model-based iterative reconstruction (MBIR) in cystic fibrosis patients undergoing treatment with cystic fibrosis transmembrane conductance regulators (CFTR). Clin Radiol 76:393.e9-393.e17. https://doi.org/10.1016/j.crad.2020.12.003
83. Mott LS, Park J, Murray CP, et al (2012) Progression of early structural lung disease in young children with cystic fibrosis assessed using CT. Thorax 67:509–16. https://doi.org/10.1136/thoraxjnl-2011-200912
84. Nasr SZ, Gordon D, Sakmar E, et al (2006) High resolution computerized tomography of the chest and pulmonary function testing in evaluating the effect of tobramycin solution for inhalation in cystic fibrosis patients. Pediatr Pulmonol 41:1129–1137. https://doi.org/10.1002/ppul.20447
85. Newbegin K, Pilkington K, Shanthikumar S, Ranganathan S (2018) Clinical utility of surveillance computed tomography scans in infants with cystic fibrosis. Pediatr Pulmonol 53:1387–1390. https://doi.org/10.1002/ppul.24132
86. Newman B, Krane EJ, Gawande R, et al (2014) Chest CT in children: anesthesia and atelectasis. Pediatr Radiol 44:164–172. https://doi.org/10.1007/s00247-013-2800-4
87. Nissenbaum C, Davies G, Horsley A, Davies JC (2020) Monitoring early stage lung disease in cystic fibrosis. Curr Opin Pulm Med 26:671–678. https://doi.org/10.1097/MCP.0000000000000732
88. Nyilas S, Bauman G, Pusterla O, et al (2019) Ventilation and perfusion assessed by functional MRI in children with CF: reproducibility in comparison to lung function. J Cyst Fibros 18:543–550. https://doi.org/10.1016/j.jcf.2018.10.003
89. Nyilas S, Bauman G, Sommer G, et al (2017) Novel magnetic resonance technique for functional imaging of cystic fibrosis lung disease. Eur Respir J 50:1701464. https://doi.org/10.1183/13993003.01464-2017
90. O'Connell OJ, McWilliams S, McGarrigle A, et al (2012) Radiologic imaging in cystic fibrosis: cumulative effective dose and changing trends over 2 decades. Chest 141:1575–1583. https://doi.org/10.1378/chest.11-1972

91. O'Connor OJ, Vandeleur M, McGarrigle AM, et al (2010) Development of Low-Dose Protocols for Thin-Section CT Assessment of Cystic Fibrosis in Pediatric Patients. Radiology 257:820–829. https://doi.org/10.1148/radiol.10100278
92. Otjen JP, Swanson JO, Oron A, et al. (2018) Spirometry-Assisted High Resolution Chest Computed Tomography in Children: Is it Worth the Effort? Curr Probl Diagn Radiol 47:14–18. doi: 10.1067/j.cpradiol.2017.02.010
93. Oudraad MCJ, Kuo W, Rosenow T, et al (2020) Assessment of early lung disease in young children with CF: A comparison between pressure-controlled and free-breathing chest computed tomography. Pediatr Pulmonol 55:1161–1168. https://doi.org/10.1002/ppul.24702
94. Owens CM, Aurora P, Stanojevic S, et al (2011) Lung Clearance Index and HRCT are complementary markers of lung abnormalities in young children with CF. Thorax 66:481–488. https://doi.org/10.1136/thx.2010.150375
95. Parad RB (2018) Non-sedation of the neonate for radiologic procedures. Pediatr Radiol 48:524–530. https://doi.org/10.1007/s00247-017-4002-y
96. Peixoto AO, Marson F AL, Dertkigil SS, et al (2020) The Use of Ultrasound as a Tool to Evaluate Pulmonary Disease in Cystic Fibrosis. Respir Care 65:293–303. https://doi.org/10.4187/respcare.07038
97. Pennati F, Roach DJ, Clancy JP, et al (2018) Assessment of pulmonary structure-function relationships in young children and adolescents with cystic fibrosis by multivolume proton-MRI and CT. J Magn Reson Imaging 48:531–542. https://doi.org/10.1002/jmri.25978
98. Pennati F, Salito C, Borzani I, et al (2019) Quantitative multivolume proton-magnetic resonance imaging in patients with cystic fibrosis lung disease: comparison with clinical indicators. Eur Respir J 53:1702020. https://doi.org/10.1183/13993003.02020-2017
99. Perez-rovira A, Kuo W, Petersen J, et al (2016) Automatic airway-artery analysis on lung CT to quantify airway wall thickening and bronchiectasis. Med Phys 1–10
100. Puderbach M, Eichinger M (2010) The role of advanced imaging techniques in cystic fibrosis follow-up: is there a place for MRI? Pediatr Radiol 40:844–9. https://doi.org/10.1007/s00247-010-1589-7
101. Puderbach M, Eichinger M, Haeselbarth J, et al (2007) Assessment of Morphological MRI for Pulmonary Changes in Cystic Fibrosis (CF) Patients. Invest Radiol 42:715–724. https://doi.org/10.1097/RLI.0b013e318074fd81

102. Puderbach M, Eichinger M, Gahr J, et al. (2007) Proton MRI appearance of cystic fibrosis: Comparison to CT. Eur Radiol 17:716–724. doi: 10.1007/s00330-006-0373-4
103. Ram S, Hoff BA, Bell AJ, et al (2021) Improved detection of air trapping on expiratory computed tomography using deep learning. PLoS One 16:1–17. https://doi.org/10.1371/journal.pone.0248902
104. Ramsey KA, McGirr C, Stick SM, et al (2017) Effect of posture on lung ventilation distribution and associations with structure in children with cystic fibrosis. J Cyst Fibros 16:713–718. https://doi.org/10.1016/j.jcf.2017.01.013
105. Ramsey KA, Rosenow T, Turkovic L, et al (2016) Lung Clearance Index and Structural Lung Disease on Computed Tomography in Early Cystic Fibrosis. Am J Respir Crit Care Med 193:60–67. https://doi.org/10.1164/rccm.201507-1409OC
106. Refait J, Macey J, Bui S, et al (2019) CT evaluation of hyperattenuating mucus to diagnose allergic bronchopulmonary aspergillosis in the special condition of cystic fibrosis. J Cyst Fibros 18:e31–e36. https://doi.org/10.1016/j.jcf.2019.02.002
107. Renz DM, Scholz O, Böttcher J, et al (2015) Comparison Between Magnetic Resonance Imaging and Computed Tomography of the Lung in Patients With Cystic Fibrosis With Regard to Clinical, Laboratory, and Pulmonary Functional Parameters. Invest Radiol 50:733–742. https://doi.org/10.1097/RLI.0000000000000178
108. Roach DJ, Crémillieux Y, Fleck RJ, et al (2016) Ultrashort Echo-Time Magnetic Resonance Imaging Is a Sensitive Method for the Evaluation of Early Cystic Fibrosis Lung Disease. Ann Am Thorac Soc 13:1923–1931. https://doi.org/10.1513/AnnalsATS.201603-203OC
109. Robinson TE, Goris ML, Moss RB, et al (2020) Mucus plugging, air trapping, and bronchiectasis are important outcome measures in assessing progressive childhood cystic fibrosis lung disease. Pediatr Pulmonol 55:929–938. https://doi.org/10.1002/ppul.24646
110. Robinson TE, Leung AN, Northway WH, et al (2001) Spirometer-triggered high-resolution computed tomography and pulmonary function measurements during an acute exacerbation in patients with cystic fibrosis. J Pediatr 138:553–559. https://doi.org/10.1067/mpd.2001.111820
111. Robinson TE, Goris ML, Zhu HJ, et al (2005) Dornase alfa reduces air trapping in children with mild cystic fibrosis lung disease: a quantitative analysis. Chest 128:2327–35. https://doi.org/10.1378/chest.128.4.2327

112. Ronan NJ, Einarsson GG, Twomey M, et al (2018) CORK Study in Cystic Fibrosis. Chest 153:395–403. https://doi.org/10.1016/j.chest.2017.10.005

113. Rosenfeld M, Emerson J, Williams-Warren J, et al (2001) Defining a pulmonary exacerbation in cystic fibrosis. J Pediatr 139:359–365. https://doi.org/10.1067/mpd.2001.117288

114. Rosenow T, Mok LC, Turkovic L, et al (2019) The cumulative effect of inflammation and infection on structural lung disease in early cystic fibrosis. Eur Respir J 54:1801771. https://doi.org/10.1183/13993003.01771-2018

115. Rosenow T, Oudraad MCJ, Murray CP, et al (2015) PRAGMA-CF. A Quantitative Structural Lung Disease Computed Tomography Outcome in Young Children with Cystic Fibrosis. Am J Respir Crit Care Med 191:1158–1165. https://doi.org/10.1164/rccm.201501-0061OC

116. Rosenow T, Ramsey K, Turkovic L, et al (2017) Air trapping in early cystic fibrosis lung disease-Does CT tell the full story? Pediatr Pulmonol 52:1150–1156. https://doi.org/10.1002/ppul.23754

117. Rowan SA, Bradley JM, Bradbury I, et al (2014) Lung clearance index is a repeatable and sensitive indicator of radiological changes in bronchiectasis. Am J Respir Crit Care Med 189:586–592. https://doi.org/10.1164/rccm.201310-1747OC

118. Rybacka A, Karmelita-Katulska K (2016) The Role of Computed Tomography in Monitoring Patients with Cystic Fibrosis. Polish J Radiol 81:141–145. https://doi.org/10.12659/PJR.896051

119. Salamon E, Lever S, Kuo W, et al. (2017) Spirometer guided chest imaging in children: It is worth the effort! Pediatr Pulmonol 52:48–56. doi: 10.1002/ppul.23490

120. Sanders DB, Bittner RCL, Rosenfeld M, et al (2011) Pulmonary exacerbations are associated with subsequent FEV1 decline in both adults and children with cystic fibrosis. Pediatr Pulmonol 46:393–400. https://doi.org/10.1002/ppul.21374

121. Sanders DB, Li Z, Brody AS (2015) Chest Computed Tomography Predicts the Frequency of Pulmonary Exacerbations in Children with Cystic Fibrosis. Ann Am Thorac Soc 12:64–69. https://doi.org/10.1513/AnnalsATS.201407-338OC

122. Sanders DB, Li Z, Brody AS, Farrell PM (2011) Chest computed tomography scores of severity are associated with future lung disease progression in children with cystic fibrosis. Am J Respir Crit Care Med 184:816–21. https://doi.org/10.1164/rccm.201105-0816OC

123. Sandvik RM, Kongstad T, Green K, et al (2021) Prospective longitudinal association between repeated multiple breath washout measurements and computed tomography scores in children with cystic fibrosis. J Cyst Fibros 20:632–640. https://doi.org/10.1016/j.jcf.2020.09.010

124. Sasihuseyinoglu AS, Altıntaş DU, Soyupak S, et al (2019) Evaluation of high resolution computed tomography findings of cystic fibrosis. Korean J Intern Med 34:335–343. https://doi.org/10.3904/kjim.2017.287

125. Schaefer JF, Hector A, Schmidt K, et al (2018) A semiquantitative MRI-Score can predict loss of lung function in patients with cystic fibrosis: Preliminary results. Eur Radiol 28:74–84. https://doi.org/10.1007/s00330-017-4870-4

126. Scholz O, Denecke T, Böttcher J, et al (2017) MRI of cystic fibrosis lung manifestations: sequence evaluation and clinical outcome analysis. Clin Radiol 72:754–763. https://doi.org/10.1016/j.crad.2017.03.017

127. Shah RM, Sexauer W, Ostrum BJ, et al (1997) High-resolution CT in the acute exacerbation of cystic fibrosis: evaluation of acute findings, reversibility of those findings, and clinical correlation. Am J Roentgenol 169:375–380. https://doi.org/10.2214/ajr.169.2.9242738

128. Sheahan KP, Glynn D, Joyce S, et al (2021) Best Practices: Imaging Strategies for Reduced-Dose Chest CT in the Management of Cystic Fibrosis–Related Lung Disease. Am J Roentgenol 1–10. https://doi.org/10.2214/AJR.19.22694

129. Sheikh SI, Long FR, McCoy KS, et al (2015) Computed tomography correlates with improvement with ivacaftor in cystic fibrosis patients with G551D mutation. J Cyst Fibros 14:84–89. https://doi.org/10.1016/j.jcf.2014.06.011

130. Sileo C, Corvol H, Boelle P-Y, et al (2014) HRCT and MRI of the lung in children with cystic fibrosis: Comparison of different scoring systems. J Cyst Fibros 13:198–204. https://doi.org/10.1016/j.jcf.2013.09.003

131. Sly PD, Brennan S, Gangell C, et al (2009) Lung Disease at Diagnosis in Infants with Cystic Fibrosis Detected by Newborn Screening. Am J Respir Crit Care Med 180:146–152. https://doi.org/10.1164/rccm.200901-0069OC

132. Stahl M, Joachim C, Wielpütz MO, Mall MA (2019) Comparison of lung clearance index determined by washout of N2 and SF6 in infants and preschool children with cystic fibrosis. J Cyst Fibros 18:399–406. https://doi.org/10.1016/j.jcf.2018.11.001

133. Stahl M, Wielpütz MO, Graeber SY, et al (2016) Comparison of Lung Clearance Index and Magnetic Resonance Imaging for Assessment of Lung Disease in Children With Cystic Fibrosis. Am J Respir Crit Care Med rccm.201604-0893OC. https://doi.org/10.1164/rccm.201604-0893OC

134. Stick S, Tiddens H, Aurora P, et al (2013) Early intervention studies in infants and preschool children with cystic fibrosis: are we ready? Eur Respir J 42:527–538. https://doi.org/10.1183/09031936.00108212

135. Strzelczuk–Judka L, Wojsyk–Banaszak I, Zakrzewska A, Jończyk–Potoczna K (2019) Diagnostic value of chest ultrasound in children with cystic fibrosis – Pilot study. PLoS One 14:e0215786. https://doi.org/10.1371/journal.pone.0215786

136. Szczesniak R, Turkovic L, Andrinopoulou E-RR, Tiddens HAWM (2017) Chest imaging in cystic fibrosis studies: What counts, and can be counted? J Cyst Fibros 16:175–185. https://doi.org/10.1016/j.jcf.2016.12.008

137. Tepper LA, Caudri D, Rovira AP, et al (2016) The development of bronchiectasis on chest computed tomography in children with cystic fibrosis: can pre-stages be identified? Eur Radiol 26:4563–4569. https://doi.org/10.1007/s00330-016-4329-z

138. Tepper LA, Ciet P, Caudri D, et al (2016) Validating chest MRI to detect and monitor cystic fibrosis lung disease in a pediatric cohort. Pediatr Pulmonol 51:34–41. https://doi.org/10.1002/ppul.23328

139. Tepper LA, Utens EMWJ, Caudri D, et al (2013) Impact of bronchiectasis and trapped air on quality of life and exacerbations in cystic fibrosis. Eur Respir J 42:371–9. https://doi.org/10.1183/09031936.00137612

140. Terheggen-Lagro S, Truijens N, van Poppel N, et al (2003) Correlation of six different cystic fibrosis chest radiograph scoring systems with clinical parameters. Pediatr Pulmonol 35:441–445. https://doi.org/10.1002/ppul.10280

141. Teufel M, Ketelsen D, Fleischer S, et al (2013) Comparison between High-Resolution CT and MRI Using a Very Short Echo Time in Patients with Cystic Fibrosis with Extra Focus on Mosaic Attenuation. Respiration 86:302–311. https://doi.org/10.1159/000343085

142. Thia LP, Calder A, Stocks J, et al (2014) Is chest CT useful in newborn screened infants with cystic fibrosis at 1 year of age? Thorax 69:320–327. https://doi.org/10.1136/thoraxjnl-2013-204176

143. Tiddens HAWM, Andrinopoulou E-R, McIntosh J, et al (2020) Chest computed tomography outcomes in a randomized clinical trial in cystic fibrosis: Lessons learned from the first ataluren phase 3 study. PLoS One 15:e0240898. https://doi.org/10.1371/journal.pone.0240898

144. Tiddens HAWM, Rosenow T (2014) What did we learn from two decades of chest computed tomography in cystic fibrosis? Pediatr Radiol 44:1490–1495. https://doi.org/10.1007/s00247-014-2964-6

145. Tiddens HAWM, Stick SM, Wild JM, et al (2015) Respiratory tract exacerbations revisited: Ventilation, inflammation, perfusion, and structure (VIPS) monitoring to redefine treatment. Pediatr Pulmonol 50:S57–S65. https://doi.org/10.1002/ppul.23266

146. Triphan SMF, Biederer J, Burmester K, et al (2018) Design and application of an MR reference phantom for multicentre lung imaging trials. PLoS One 13:e0199148. https://doi.org/10.1371/journal.pone.0199148

147. Triphan SMF, Stahl M, Jobst BJ, et al (2020) Echo Time-Dependence of Observed Lung T1 in Patients With Cystic Fibrosis and Correlation With Clinical Metrics. J Magn Reson Imaging 52:1645–1654. https://doi.org/10.1002/jmri.27271

148. Turkovic L, Caudri D, Rosenow T, et al (2020) Structural determinants of long-term functional outcomes in young children with cystic fibrosis. Eur Respir J 55:1900748. https://doi.org/10.1183/13993003.00748-2019

149. van Straten M, Brody AS, Ernst C, et al (2020) Guidance for computed tomography (CT) imaging of the lungs for patients with cystic fibrosis (CF) in research studies. J Cyst Fibros 19:176–183. https://doi.org/10.1016/j.jcf.2019.09.001

150. VandenBranden SL, McMullen A, Schechter MS, et al (2012) Lung function decline from adolescence to young adulthood in cystic fibrosis. Pediatr Pulmonol 47:135–143. https://doi.org/10.1002/ppul.21526

151. Verbanck S, King GG, Zhou W, et al (2018) The quantitative link of lung clearance index to bronchial segments affected by bronchiectasis. Thorax 73:82–84. https://doi.org/10.1136/thoraxjnl-2017-210496

152. Verbanck S, Vanderhelst E (2018) The Respective Roles of Lung Clearance Index and Magnetic Resonance Imaging in the Clinical Management of Patients with Cystic Fibrosis. Am J Respir Crit Care Med 197:409–409. https://doi.org/10.1164/rccm.201706-1137LE

153. Voskrebenzev A, Gutberlet M, Klimeš F, et al. (2018) Feasibility of quantitative regional ventilation and perfusion mapping with phase-resolved functional lung (PREFUL) MRI in healthy volunteers and COPD, CTEPH, and CF patients. Magn Reson Med 79:2306–2314. doi: 10.1002/mrm.26893

154. Vult von Steyern K, Björkman-Burtscher IM, Geijer M (2013) Radiography, tomosynthesis, CT and MRI in the evaluation of pulmonary cystic fibrosis: an untangling review of the multitude of scoring systems. Insights Imaging 4:787–798. https://doi.org/10.1007/s13244-013-0288-y

155. Wagener JS, Elkin EP, Pasta DJ, et al (2015) Pulmonary function outcomes for assessing cystic fibrosis care. J Cyst Fibros 14:376–83. https://doi.org/10.1016/j.jcf.2014.11.008

156. Weinheimer O, Hoff BA, Fortuna AB, et al (2019) Influence of Inspiratory/Expiratory CT Registration on Quantitative Air Trapping. Acad Radiol 26:1202–1214. https://doi.org/10.1016/j.acra.2018.11.001

157. Wielpütz MO, Eichinger M, Puderbach M (2013) Magnetic Resonance Imaging of Cystic Fibrosis Lung Disease. J Thorac Imaging 28:151–159. https://doi.org/10.1097/RTI.0b013e31828d40d4

158. Wielpütz MO, Eichinger M, Wege S, et al (2019) Midterm Reproducibility of Chest Magnetic Resonance Imaging in Adults with Clinically Stable Cystic Fibrosis and Chronic Obstructive Pulmonary Disease. Am J Respir Crit Care Med 200:103–107. https://doi.org/10.1164/rccm.201812-2356LE

159. Wielpütz MO, Eichinger M, Weinheimer O, et al (2013) Automatic Airway Analysis on Multidetector Computed Tomography in Cystic Fibrosis. J Thorac Imaging 28:104–113. https://doi.org/10.1097/RTI.0b013e3182765785

160. Wielpütz MO, Puderbach M, Kopp-Schneider A, et al (2014) Magnetic Resonance Imaging Detects Changes in Structure and Perfusion, and Response to Therapy in Early Cystic Fibrosis Lung Disease. Am J Respir Crit Care Med 189:956–965. https://doi.org/10.1164/rccm.201309-1659OC

161. Wielpütz MO, von Stackelberg O, Stahl M, et al (2018) Multicentre standardisation of chest MRI as radiation-free outcome measure of lung disease in young children with cystic fibrosis. J Cyst Fibros 17:518–527. https://doi.org/10.1016/j.jcf.2018.05.003

162. Wijker NE, Vidmar S, Grimwood K, et al (2020) Early markers of cystic fibrosis structural lung disease: follow-up of the ACFBAL cohort. Eur Respir J 55:1901694. https://doi.org/10.1183/13993003.01694-2019

163. Willmering MM, Roach DJ, Kramer EL, et al (2021) Sensitive structural and functional measurements and 1-year pulmonary outcomes in pediatric cystic fibrosis. J Cyst Fibros 20:533–539. https://doi.org/10.1016/j.jcf.2020.11.019

164. Woods JC, Wild JM, Wielpütz MO, et al (2020) Current state of the art MRI for the longitudinal assessment of cystic fibrosis. J Magn Reson Imaging 52:1306–1320. https://doi.org/10.1002/jmri.27030

165. Yammine S, Ramsey KA, Skoric B, et al (2019) Single-breath washout and association with structural lung disease in children with cystic fibrosis. Pediatr Pulmonol 54:587–594. https://doi.org/10.1002/ppul.24271

166. Zorzo C, Caballero P, Diab L, et al (2020) Predictive value of computed tomography scoring systems evolution in adults with cystic fibrosis. Eur Radiol 30:3634–3640. https://doi.org/10.1007/s00330-020-06759-z

167. Zucker EJ, Barnes ZA, Lungren MP, et al (2020) Deep learning to automate Brasfield chest radiographic scoring for cystic fibrosis. J Cyst Fibros 19:131–138. https://doi.org/10.1016/j.jcf.2019.04.016

Table 2E Radiologist-Clinician cooperation:

Statement number	Statement	Type of statement	Strength of recommendation	Quality of evidence	Most relevant supporting articles
2E.1	There are no studies addressing optimal collaboration strategies between clinicians and radiologists as members of CF multidisciplinary teams. Research should be addressed to answer this topic	Statement of fact		Best Practice	n/a
2E.2	At least a radiologist, with experience in CF lung disease, should be part of the CF multidisciplinary team. This radiologist should periodically attend multidisciplinary team meetings to discuss CF patients	Recommendation	Grade A	Best Practice	n/a

n/a=not available.

Table 3E. Structured report:

Statement number	Statement	Type of statement	Strength of recommendation	Quality of evidence	Most relevant supporting articles
3E.1	More research is needed to fully understand the benefits of STR (Structured Radiological Report) for CF patients	Statement of fact		Best Practice	n/a
3E.2	STR specific for CF should be validated for use in clinical practice and as research outcome	Recommendation	Grade A	Best Practice	n/a

n/a=not available.

Table 4E. Statements related to imaging modality

Statement number	Statement	Type of statement	Strength of recommendation	Quality of evidence	Most relevant supporting articles
Computed tomography (CT)					
4E.1	Spirometry assisted CT improves lung volume standardization and air trapping detection, but no appreciable difference in image quality is detected in terms of presence of motion artifact or atelectasis, especially in inspiratory scan. Therefore, in the appropriate clinical setting spirometry controlled CT is favourable for CF imaging in cooperative patients, but not mandatory	Recommendation	Grade B	Moderate	[Online supplement reference list 91,118]
4E.2	In uncooperative children (younger than 6 years), CTs can be acquired during free breathing with fast multi-slice CT-scanners, in order to avoid risks related to general anaesthesia and sedation.	Recommendation	Grade B	High	[Online supplement reference list 64, 92]
Magnetic Resonance imaging(MRI)					
4.E3	Image quality of MRI varies between MR brands and full clinical implementation will require further development and validation work through multicentre standardization to support its use as radiation-free outcome measure of lung disease.	Recommendation	Grade C	High	[Online supplement reference list 145, 160]
4E.4	To maximize the potential of CT and MRI markers in clinical studies and advance treatment of CF disease progression, efforts should be made to develop data repositories, promote standardization and conduct reproducible research.	Recommendation	Grade I	Moderate	[Online supplement reference list 135]

4E.5	Inflammation MRI A. Quantification of volume of high T2-weighted signal is a promising quantitative outcome measure of lung inflammation B. Diffusion weighted Magnetic Resonance Imaging (DWI-MRI) is a promising imaging method for non-invasive detection of pulmonary inflammation during pulmonary exacerbation, and might be used to monitor treatment efficacy of anti-inflammatory treatment.	Statement of fact	///	Low	[Online supplement reference list 8, 24, 25]
4.E6	Ventilation/Perfusion MRI A. SI and lung volumes are potential MRI quantitative parameters to detect lung impairment, as expressed by low intensity region B. Fourier Decomposition MRI is a reliable quantitative parameter of ventilation (and perfusion) in CF lung disease C. Hyperpolarized gas quantitative ventilation MRI parameters correlate with amount of trapped air in early CF lung disease as expressed by lung function and can have an impact on therapy management. D. Ventilation MRI with Fluorinated gas technology is not currently as well developed as hyperpolarized gas MRI, but there is a strong potential for using it in similar	Statement of fact	///	Moderate	[Online supplement reference list 9, 45, 52, 102, 110] And [Online supplement reference 31, 49, 50, 152]

	applications in the future E. Limited post-processing tool hamper translation of this techniques in clinical practice.				