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Guidance for chest-CT in children and adults with cystic fibrosis: A European perspective

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ABSTRACT

The European Cystic Fibrosis Society-Clinical Trials Network (ECFS-CTN) herein proposes guidance for the use of chest CT-scans for the regular monitoring of lung disease in CF. Statements were completed in a 3-step process: the questions were identified via an anonymous online survey, followed by a comprehensive literature search, and a final Delphi process. The guidance recommends the use of ultra-low dose CT scans (effective radiation dose, 0.08 mSv; equivalent to 2 to 4 chest X-rays), tracking of patients' cumulative radiation and effective communication strategies using "de-medicalized" information for shared decision making. Chest CT scans (with lung volume monitoring) are not recommended systematically in both children and adults. Ultimate responsibility for justifying a chest CT scan lies with the individual professionals directly involved, the final decision being influenced by indications, costs, expertise, available material, resources and/or the patient's values, as well as possible impact on treatment modalities.

1. Introduction

Thoracic imaging in cystic fibrosis (CF) has become increasingly important in recent years as it allows direct visualisation of structural lung disease at a very early age [1]. The reluctance to use these techniques more widely is due to many factors such as the choice of method used (conventional chest radiography, CT scan, magnetic resonance imaging of the lung (MRI), etc.), the type of strategy used to reduce the CT radiation dose [2], the possible need for sedation and the importance of these techniques compared to spirometry and measurement of the lung clearance index (LCI) [3]. The peculiarities of imaging in adults compared to children must also be considered. Although the technique of chest CT is not new, it has evolved over time [1,2,4], and in the era of more modern devices, CF transmembrane conductance regulator (CFTR) modulators, and the advent of lung MRI, the use of chest CT has changed dramatically [3].

Some studies have evaluated the use of chest CT scans in CF and suggested priorities for their use in people with CF (PwCF). In one study, responses were obtained from 27 clinical leads and 22 senior radiographers in the UK [5]. Ninety-three percent of clinicians reported that chest CT was useful in monitoring disease progression and 70 % said it often changed management. However, only 5 (19 %) performed routine scans. When indicated, 6 (27 %) used sedation and 16 (73 %) used general anaesthesia. Only 1 (5 %) routinely used intravenous contrast and 3 (14 %) routinely took expiratory images [5].

A consortium of 21 experts in pulmonology and radiology in CF from the ten largest specialist centres in Italy presented an overview of the state of lung imaging in CF with recommendations for the "iMAging managEment of cySTic fibROsis" (MAESTRO) [6]. Although rigorous PICO (patient/population, intervention, comparison and outcomes)-style responses were provided, firm recommendations could not be made for some important clinical situations. [6]. It was concluded that international guidance is needed on the appropriate timing and choice of imaging modalities for patients with CF lung disease depending on the clinical situation, patient age, lung function and type of treatment.

Here we propose practical European guidance based on 1) 2 online surveys by the European CF Society Clinical Trials Network (ECFS CTN), 2) the available literature, and 3) a Delphi method of validation to produce such guidance [8].

2. Methodology

Our guidance is based on a careful review of the literature and on surveys and consensus discussions with experts in the field. A multidisciplinary committee of 7 pediatric pulmonologists, 5 adult radiologists, 5 pulmonologists, 1 standardization specialist, 1 federal patient organisation representative, all experienced in CF care, and a study methodologist were involved in overseeing this work.

An initial survey to identify the most important issues on the subject was sent to all 57 ECFS CTN CF centres and the members of the ERS CF Task Force. A total of 43 responses were received for the pediatric questionnaire and 22 for the adult questionnaire, mainly from CF pulmonologists, but also from radiologists (9 and 4 in the questionnaires, respectively). Questions, in PICO format where appropriate, were elaborated.

Databases were then searched, using PubMed, Cochrane Library, Scopus, Embase and Web of Science. The reference lists of all included articles were also searched for additional articles of interest. Where appropriate, more detailed search strategies are provided in the subsections of this manuscript. The authors considered original research, randomized controlled trials, systematic reviews or meta-analyzes, guidelines and consensus statements. Articles and case reports not written in English were excluded. The search was conducted regardless of the date of the study. Relevant papers were ranked, and the level of evidence was graded according to the United States Preventative Services Task Force (USPFTF) system for assessing the quality of evidence and strength of recommendations. In this document, systematic analyses were not considered possible due to the lack of homogeneous studies.

The committee drew up a list of guidance proposals based on either evidence or expert opinion. To counter possible bias from selection of key issues from the initial survey of pulmonologists and radiologists, the Delphi rounds were then used to reach agreement or disagreement amongst a wider group of $>$20 specialists in CF from ECFS CTN paediatric and adult specialist centres including radiologists, medical physicists, allied health professionals and pulmonologists, voting online using an anonymous electronic voting system via an encrypted ePlatform and meeting via Zoom for discussion of each subsequent Delphi round of voting.

Participants voted using a nine-point Likert scale: strongly disagree (score 1), disagree (score 3), neutral (score 5), agree (score 7) and strongly agree (score 9) [9]. In accordance with methodology recommendations we excluded a maximum of one extreme value per 15 members if no value was missing [9], e.g. the minimal value if the median was $>$5 or the maximal value if the median was $\leq$5). Rounds were ended when consensus was reached or when it was clear that no agreement could be reached: stable disagreement $\geq$2 rounds or when a maximum of 3 rounds were completed [10]. Round 1, round 2 and round 3 comprised 19, 14 and 24 participants, respectively. Voting outcomes for each Delphi statement are reported in individual tables for each statement.

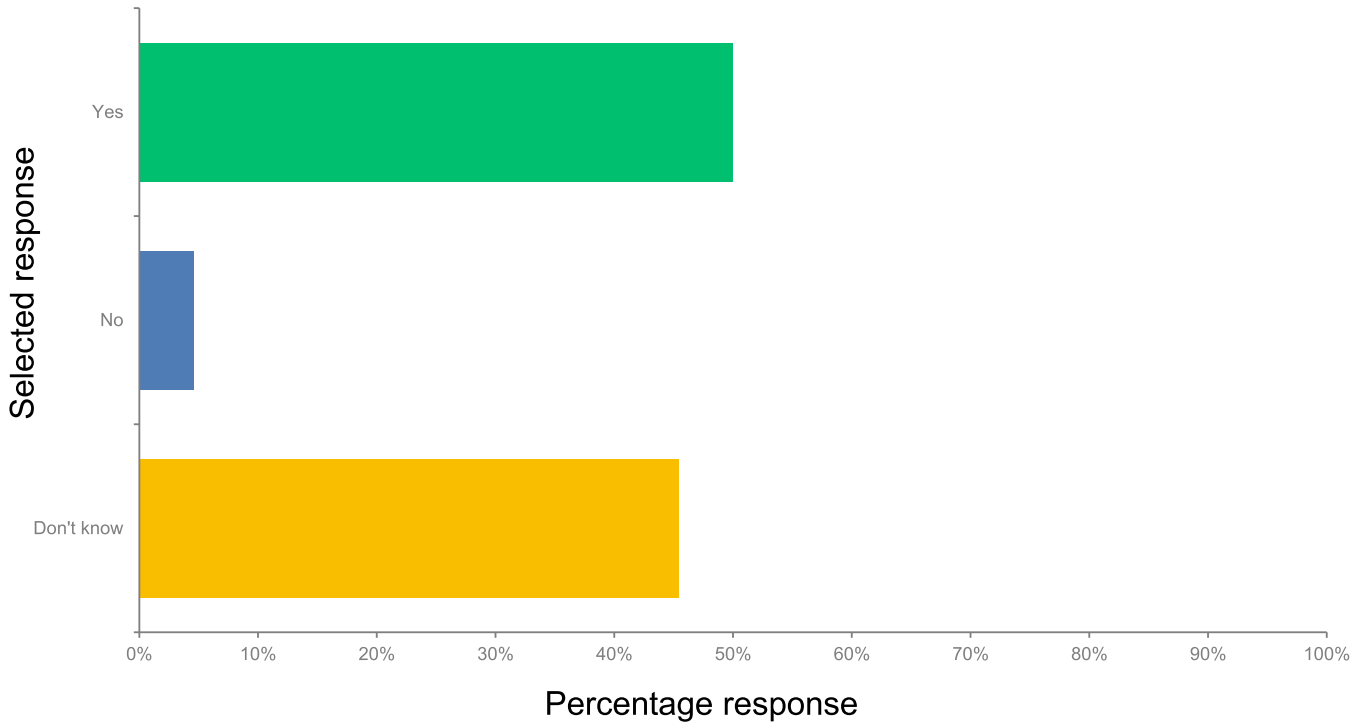

Fig. 1. Availability of low-dose CT scans.

3. Radiation dose

3.1. From the patient perspective

Informing the patient about radiation exposure in connection with an imaging examination is now a mandatory part of the radiology procedure and contributes to the relationship of trust between the patient and their caregiver. The European Union Council Directive states that "*Information on the patient's radiation exposure shall be part of the report on the medical radiological procedure.*" There is much debate about the optimal method of communicating such information, with recent research showing that patients are widely under-informed about the risks of medical radiation and do not understand the terminology for imaging procedures [7,8]. The efforts of all healthcare professionals to develop an effective communication strategy are crucial [9]. Related to the concept of a unified message to patients, there is a need to develop more user-friendly, patient-centered educational materials that are not written at a higher than recommended reading level [10–12], and to consider modified language when communicating concepts related to the complex issue of radiation exposure and associated risks [13]. "*What is needed is a systemic and seismic shift in educating physicians and patients, in having candid conversations with patients around imaging that acknowledge the trade-offs, and in justifying the use of all medical radiation exposure. In doing so, we improve the safety of medical imaging while reducing the physical, social, and economic toll of overuse and disease.*" [14].

There has been much discussion about the optimal content of patient-friendly information, ranging from communicating the extent of radiation exposure of an imaging examination in terms of its equivalence to that of a CXR, to the time for equivalent background radiation exposure or comparison with international flight times [15,16] (see section 3.4 below).

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
1	The communication of radiation dose and associated risks must be done through an effective communication strategy and concrete, "de-medicalized" information so that both patients and physicians can develop a better understanding and improve shared decision-making	Grade D*	4	9 [6–9] APPROPRIATE (Relatively Strong Agreement)

*RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

3.2. Risk-benefit analysis of sequential chest-CT in different dose levels and age groups

Sequential chest-CT continues to be used in certain facilities because it can limit radiation dose by acquiring incremental images with gaps and no overlap between slices [17,18]. However, the use of sequential CT is limited by the possibility of missed pathology and may impact the visualisation and characterization of disease that spreads

craniocaudally, i.e., outside the axial imaging plane (e.g., airway pathology) and is therefore incompletely visualised in a single axial image, potentially problematic in longitudinal follow-up. Furthermore, the protocol cannot be readily used with manual or automated scoring systems [19,20]. For patients who have difficulty holding their breath or remaining still during the scan, such as young children or patients with movement disorders, sequential imaging may be preferable to a helical protocol to avoid motion artefacts.

Advances in CT technology and the implementation of dose optimization techniques, such as tube current modulation, kilovoltage peak (kVp) selection, organ-based dose modulation, and advanced image reconstruction algorithms, such as iterative reconstruction, have enabled the development of low-dose and ultra-low-dose CT protocols [1,21]. These protocols aim to minimize radiation exposure without significantly compromising diagnostic image quality. Although the benefit-risk ratio of low-dose and ultra-low-dose CT is generally favourable, the diagnostic accuracy and clinical utility of these protocols must be carefully evaluated on a case-by-case basis, taking into account factors such as the patient's age, clinical presentation, underlying disease, availability of previous imaging studies, and most importantly, the underlying clinical question or specific diagnostic task [1,6,22].

3.3. Definition of the low dose

In the online survey, half of the respondents indicated that ultra-low dose chest CT was "available" in their centres (Fig. 1). Of note, the definition of ultra-low dose was not provided, nor were the clinical indications for the use of these protocols or whether they are used in specific patient groups.

There is no universal definition of a "low dose" or "ultra-low dose" in CT, as dose can vary by country and change over time with technological advances resulting from the continuous efforts of radiologists and physicists to optimise dose [23]. Previous studies have used different dose values and measurements, including effective dose (ED) and size-specific dose estimation (SSDE), making it difficult to compare results [24,23,25]. The advent of new CT technologies, including image reconstruction algorithms, tube potential optimization, and noise reduction algorithms, has accelerated this dose reduction [26–28], resulting in protocols previously considered low dose becoming the accepted standard dose [23]. With the rapid pace of technological advancement, the meaning of today's term "low dose" will become even more ephemeral and will likely become obsolete.

ED, measured in millisieverts (mSv) (1 mSv = 100 mrem = 1 mGy), a calculated value that cannot be measured directly [24], is based on the hypothetical relative radiosensitivity of the organs exposed during a given CT scan protocol [21,24]. The International Commission on Radiological Protection (ICRP) defines weighting factors that are assigned to these organs based on a "reference human" that is regularly re-evaluated [29] (ED = DLP (dose-length product) x weighting factor). DLP considers the dose per slice and the length of the scan as well as the stochastic risk for an examination type covering the same anatomical region. For chest CT, the weighting factor is 0.039, 0.026, 0.018, 0.013 and 0.014 for 0, 1, 5, 10-year-old children and adults respectively [30,31].

The typical ED of a standard dose chest CT has been reported as 3–8 mSv [32], while a low dose study is typically described as less than 1 mSv [33] and the ultra-low dose is approximately 0.08 mSv [34]. With the increasing availability of modern CT scanners with improved technology to optimise radiation dose, conventional CT thorax has recently been performed at lower CT radiation doses approaching 1–1.7 mSv, although radiation dose varies by geographic location [32,35,36].

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
2	The typical effective dose of a single standard dose chest- CT scan should be in the range of 1.5–8 mSv, a low-dose scan should typically be less than 1 mSv, and an ultra-low-dose approximately 0.08 mSv (equivalent to 1.5 to 4 PA [posterior-anterior] chest radiographs).	Grade C*	3	9 [5–9] APPROPRIATE (Relatively Strong Agreement)

*RECOMMENDATION OF MODERATE CERTAINTY.

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
3	Performing ultra-low-dose CT scans in every institution should be considered a priority in the management of PwCF	GRADE D*	4	9 [5–9] APPROPRIATE (Relatively Strong Agreement)

*RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
4	In PwCF in whom the main goal of the CT scan is to evaluate the bronchi (bronchiectasis), bronchioles and lung parenchyma for ground-glass or consolidation changes (without a thorough examination of the peripheral interstitium), low-dose and ultra-low-dose CT techniques are adequate.	Grade C*	2+	9 [7–9] APPROPRIATE (Strong Agreement)

*RECOMMENDATION OF MODERATE CERTAINTY.

However, the effective dose is unsuitable to describe the dose for a particular individual, mainly because it does not consider the influence of a patient's size on the dose and because the organ coefficients are averaged over age groups and genders. To assess the risk for a particular individual, it is best to consider the specific organ doses received by the patient [23,37,38].

3.4. Communicating imaging tests and their equivalence (comparators)

When communicating the risk associated with ionising radiation, it is common to use a comparator to put the radiation dose into perspective. The choice of comparators can have a significant impact on how people perceive the risk associated with a particular exposure.

A common approach is to compare the radiation exposure from a particular imaging study with the radiation exposure that humans naturally absorb from the environment, often referred to as background radiation. The average background radiation is reported to be around 3 mSv per year [39], although this can vary depending on geographical location, altitude, and other factors.

In addition, healthcare providers may wish to use multiple comparators or different formulations to provide patients with a better understanding of the risks associated with medical radiation exposure. For example, they may compare the radiation dose from a medical imaging study with the dose they receive from other everyday activities, such as flying on an airplane (a transatlantic flight = 0.08 mSv) or the effects of ultraviolet radiation from sun exposure [40]. This can help to provide a more comprehensive picture of the risks associated with medical radiation exposure and make it easier for patients to make informed decisions about their healthcare.

3.5. How can we reduce patient anxiety levels?

The experience of undergoing medical imaging procedures, including CT scans, can cause a high level of anxiety in some patients. A number of factors contribute to patient anxiety during such a diagnostic procedure [41]. These include fears about potential diagnostic findings or possible changes in therapeutic treatment, being in an unfamiliar environment, the need to remain still for a period of time during the scan, the acoustic disturbances associated with the scan, communication difficulties during the procedure and concerns related to radiation exposure [42]. The use of a sedative poses potential risks to the patient, complicates the scanning process and prolongs the patient's stay in the radiology department [43].

Numerous strategies can be implemented to alleviate patient anxiety both prior to and during CT examinations. A cornerstone of such anxiety management is thorough patient education. The role of the radiographer goes beyond image acquisition; continuous reassurance and regular information about the progress of the examination contributes to a more personal, patient-centered experience [44]. This consistent communication can transform the procedure from an intimidating clinical process into a manageable, less stressful event for the patient. Studies assessing anxiety during MR imaging, which is more commonly associated with claustrophobia, have shown favourable reductions in anxiety scores through the use of relaxation techniques [45], music [46], audio-visual systems [47] and anxiolytics [43]. If institutional guidelines allow, a familiar person accompanying the patient during the procedure can provide essential emotional reassurance, especially in paediatric patients or those with cognitive impairment.

3.6. Impact of radiation passport or radiation dose monitoring software/tracking systems on radiation dose in CF

PwCF often require recurrent imaging, particularly chest X-rays and CT of the abdomen and thorax, to monitor the progression of their disease, detect acute illness or disease exacerbations, and make treatment decisions [24]. Over time, repeated imaging can result in a significant cumulative radiation dose, which may raise concerns about lifetime malignancy risk [24,48]. To address this issue, large multinational companies with expertise in medical imaging have developed radiation dose monitoring software or tracking systems that monitor and manage all aspects of radiation exposure associated with medical imaging.

Currently, many countries have adopted radiation tracking as standard practice [49], and there are systems that can also record a patient's radiation exposure history and are able to convert the radiation dose from the imaging procedure into an effective dose and perform a calculation to determine the cumulative effective dose (CED) [50]. An International Atomic Energy Agency-supported multinational study evaluating CED data from patients with chronic conditions requiring recurrent imaging recommended the establishment of cumulative radiation exposure thresholds in dose management systems to alert healthcare providers to potential risks to patients who have previously received high cumulative doses [50]. Previous studies have shown better justification and optimization of radiation protection by tracking individual patient doses [49,51]. National radiation dose tracking systems integrated into the electronic medical record [49,52] or a personal radiation passport [53] would provide a more accurate representation of the patient's CED and further improve physicians' access to this relevant information.

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
5	A national tracking system (or personal radiation passport) integrated into the electronic patient record to monitor cumulative radiation dose, to establish cumulative dose thresholds and to better justify and optimise radiation protection at PwCF is warranted	Grade B*	2++	9 [7–9)] APPROPRIATE (Strong agreement)

*RECOMMENDATION OF MODERATE TO HIGH CERTAINTY.

4. Early structural lung disease and early interventions

4.1. Physiopathology of early lung disease

Observational data from infant and animal models show that CFTR dysfunction in the airways can be present from the first days of life, causing a muco-inflammatory milieu with dehydrated airway surface fluid and abnormal mucin secretion. This leads to airway obstruction, which favours early infection and triggers inflammation [54] leading to bronchiectasis.

In light of these data, screening for early lung injury is a key process in the clinical management of PwCF. However, the extensive use of invasive methods such as flexible bronchoscopy and bronchoalveolar lavage is not always feasible both in routine clinical practice and as an endpoint of studies. In addition, spirometry alone is not considered sensitive enough for detecting early changes, especially in young PwCF, whose participation might be inadequate [55].

4.2. CT features of early CF lung disease

Our understanding of early lung injury in neonates and preschool children with CF (CwCF) has been enhanced by several research studies using flexible bronchoscopy and chest CT scans as clinical endpoints [56–58]. While bronchoalveolar lavage (BAL) detects neutrophilic inflammation, high-resolution chest CT (HRCT) has been shown to be a sensitive method to detect early structural changes, even without

clinical signs of airway manifestations. Thickening of the airway walls and air trapping were the most common radiological changes in this group. In a cornerstone series of studies conducted by the Australian AREST-CF team, bronchial wall thickening was found in approximately 45 % and air trapping in approximately 66 % of infants with CF at 3 months of age diagnosed by newborn screening (NBS), the majority of whom had no clinical signs of CF lung disease at the time of investigation [56].

The improvement in radiological scores following treatment of pulmonary exacerbations suggests that early structural changes may be reversible, at least to some degree [59]. However, a recent longitudinal study using annual MRI scans showed progression of early CF lung disease in the first four years of life under symptomatic therapy [60]. Children diagnosed with NBS had milder structural CF lung disease in the first years of life than clinically diagnosed infants and preschool children, but the annual rate of progression was comparable between these groups, emphasising the need for more effective therapies to prevent or slow progression [60], such as CFTR modulator therapy.

4.3. Effects of early interventions on structural changes in thoracic CT

The early initiation of appropriate preventive and curative treatments, carried out in specialised CF centres, is crucial to reduce the severity of the disease. In terms of lung disease, it remains a challenge to prevent irreversible lung damage due to lung infections and inflammation that develop in CwCF early in childhood [61]. In 2005, Robinson et al. studied the effect of DNAse on respiratory function and anatomical damage on chest CT images in 25 children with CF who were treated for one year. After three months, the treated group showed a 13 % decrease in the presence of air trapping at T0 compared to the untreated group, which showed a 48 % increase in the same parameter [62].

Studies with twice-daily inhaled hypertonic saline (7 % NaCl) have shown an improvement in lung clearance index and structural lung changes in preschool children compared to children treated with isotonic saline, as measured by chest CT after 48 weeks of treatment [63, 64]. Total lung disease (%Disease) was approximately 57 % higher in the isotonic saline (IS) group than in the hypertonic saline (HTS) group after 48 weeks. This was mainly due to the progression of bronchiectasis (1.20 % (0.93–1.47) in the IS group versus 0.72 % (0.45–0.99)) in the HTS group ($p = 0.021$)) and air trapping (2.94 % (2.30–3.59) in the IS group and 0.90 % (0.33–1.47) in the HTS group ($p = 0.0063$). This suggests that inhaled HTS should be introduced as maintenance therapy in early childhood.

The COMBAT study, in which oral azithromycin was administered to infants from the time of diagnosis of CF after NBS, showed a reduction in airway inflammation and morbidity. Improvements in clinical outcomes included reduced days hospitalised for pulmonary exacerbations, and days of hospitalisation in the first year of life, reduced inhaled or oral antibiotics, and decreased concentrations of airway inflammation biomarkers in the BAL (IL-8 and neutrophil elastase activity at 36 months of age) [65]. However, azithromycin treatment had no effect on the primary outcomes of structural lung disease, with Perth-Rotterdam Annotated Grid Morphometric Analysis-CF (PRAGMA-CF) scores showing a similar degree of bronchiectasis at 36 months (active treatment group (88 %) versus placebo (94 %)). These results suggest that thrice-weekly azithromycin can be considered for routine early treatment of CwCF, but that its effects on structural lung disease remain to be demonstrated.

New CFTR modulator therapies that correct the underlying molecular defect are available at a young age. These treatments give great hope that this severe and life-threatening disease can finally be brought under control at an early age. A preliminary real-life retrospective study including a few preschool children showed that CFTR modulators improve several quantitative CT outcomes [66]. This study included 16 modulator-exposed PwCF and 25 unexposed PwCF. Median (range) age at baseline was 12.55 (4.25–36.49) years and 8.34 (3.47–38.29) years, respectively. PRAGMA-CF %Airway disease (−2.88 (−4.46, −1.30), $p = 0.001$) and %Bronchiectasis extent (−2.07 (−3.13, −1.02), $p < 0.001$) improved in exposed PwCF vs. unexposed patients [66].

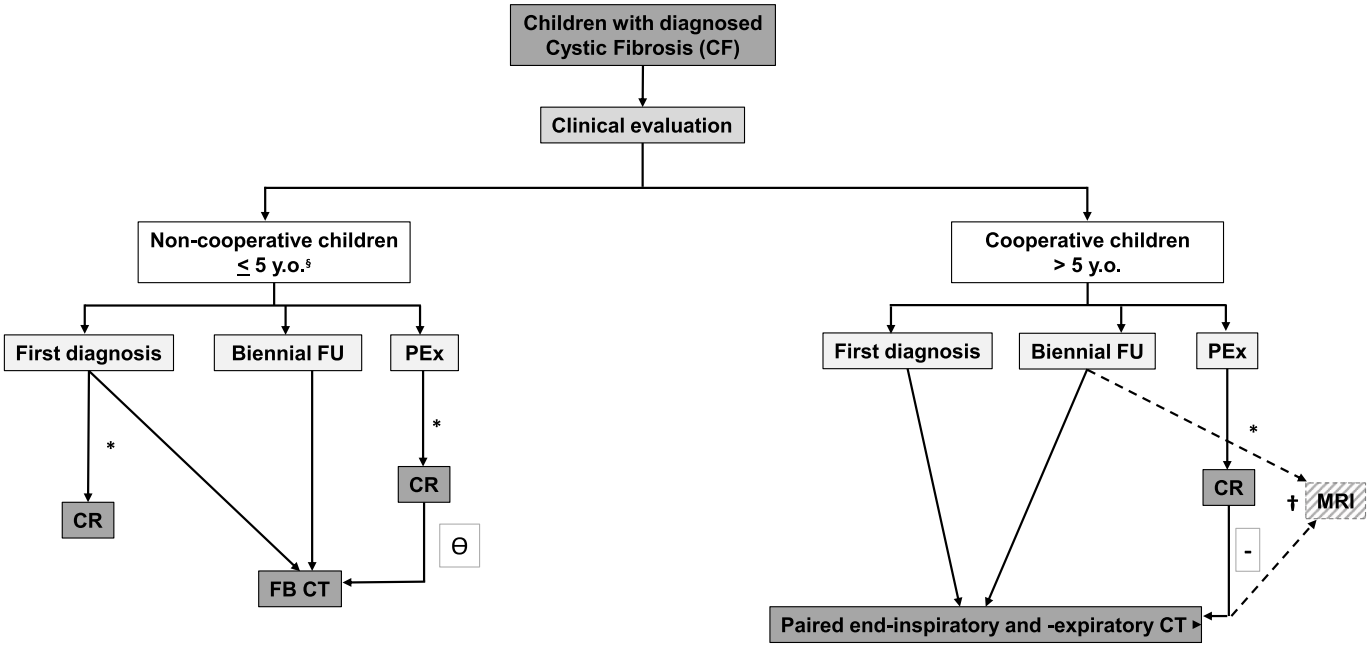

Fig. 2. Current diagnostic algorithm for cystic fibrosis (CF) imaging at the Erasmus MC-Sophia Children's Hospital, Rotterdam, The Netherlands, reproduced with permission from Ciet et al [76]. Current diagnostic algorithm for cystic fibrosis (CF) imaging. *CR* chest radiography, *CT* computed tomography, *FB* free breathing, *FU* follow-up, *MRI* magnetic resonance imaging, *PEx* pulmonary exacerbation, *y.o.* years old. The term "CT" in the flow chart is a general term, where dose should be set according to current dose reference level for paediatric chest CT imaging. § Non-cooperative children are generally those who are younger than 5 years or have developmental disabilities, * depending on cystic fibrosis centre expertise, ⊖ CT can be performed in cases of negative chest radiography findings and persistent symptoms despite therapy, † at our centre we use MRI for short-term follow-up of pulmonary exacerbation and as follow-up technique in stable CF subjects in alternation with CT (1 year CT and the year after MRI).

5. Indications, timing and frequency of CT imaging

5.1. In children

5.1.1. Initial CT scan examination in very young infants

In a recent ECFS CTN survey, 63 % of centres performed an initial CT scan of the chest, mostly at the age of 5 years. In CwCF aged less than 1 year, frequent [67] structural changes detected on chest CT are generally mild [68] and poorly reproducible (Brody II scores), with the exception of air trapping [69]. Other scores such as quantitative PRAGMA-CF outcomes show high intra-observer and inter-observer agreement [70] in the majority of studies conducted in the Netherlands and Australia. However, in one study conducted in Denmark in children aged 6–18 years, although the intra-observer correlation coefficients (ICCs) were excellent for %Bx (the proportion of the lung volume with bronchiectasis), %Dis (the proportion of the lung volume with total disease) (both ICCs >0.92), and %TA (the proportion of the lung volume with trapped air) (ICC = 0.83), the inter-observer intra-class ICC for PRAGMA-CF scores were lower: %Bx 0.56, %Dis 0.75, and %TA 0.96 [71]. PRAGMA-CF is not available for general use like the Brasfield or Brody II scoring systems (in addition to special software, approval from the owner of PRAGMA-CF is required). Overall, although the risk of chest CT is low, this examination is of dubious value on an individual basis [72] and should therefore not be routinely used in very young infants (age 1 year) [68,69] if other parameters are satisfactory (body growth, no recurrent infections or exacerbations, normal LCI, etc.).

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
6	Chest CT should not be routinely used in very young infants if other parameters are satisfactory (no overt infections requiring antibiotics beyond the usual range for such infants, no exacerbations, normal lung clearance index, etc.).	GRADE D*	4	9 (5–9] APPROPRIATE (Relatively Strong Agreement)

*RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

A chest CT is justified as soon as the physician considers that the child is unwell (pancreatic insufficiency with poor growth, repeated intravenous antibiotic treatment, recurrent lower respiratory tract infections in the first three years of life and inflammation of the lower respiratory tract) [72], regardless of the patient's age, since a single CT scan is not associated with an increased risk of subsequent intracranial tumors, leukaemia or lymphoma in children [73]. In this case, a CT scan outperforms CXR (higher sensitivity of CT in detecting bronchiectasis) [6,74].

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
7	An initial chest CT is justified if the doctor believes that a child with CF is not doing well (repeated intravenous antibiotics, recurrent lower respiratory tract infections in the first three years of life and inflammation of the lower respiratory tract) and if other investigations without radiotherapy do not find a cause.	GRADE D*	4	9 [6–9] APPROPRIATE (Relatively Strong Agreement)

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(continued)

*RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

5.1.2. First CT scan in older children

In older children, the potential risks of radiotherapy are less considered [75]. In some centres, planned age-based CT scans are performed (Fig. 2).

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
8	An ***initial routine baseline CT scan*** should be considered at around 5–7 years of age, regardless of the child's clinical condition.	GRADE D*	4	8 [1–9] UNCERTAIN (No consensus)

*RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

5.1.3. Follow-up CT-scan

In the ECFS CTN survey, 67 % of centres performed *repeat* CT scans, even if the child remains healthy, mostly at three-year intervals. For children and adults with stable disease, the ECFS recommends an annual CXR [77,78]. Systematic biennial (every two years) follow-up chest CT scans have been proposed for children of all ages [76]. However, the time interval between two consecutive CT examinations must be adapted to the clinical picture. This is because the progression of bronchiectasis is different in older children: slow or rapid. Salamon et al. identified all patients in a rapid progression group as pancreatic insufficient [79], whereas this was the case in 80 % of patients in the slow progression group. Most bronchiectatic airways developed within 2 years (rapid progression between consecutive scans) without visible precursors and may be missed on biennial CT scans. Therefore, in children with marked mucus plugging (filling of clearly identifiable bronchi), an annual CT scan may be considered to detect rapid progression of bronchiectasis [80].

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
9	In preschool children, ***routine follow-up CT scans*** are not recommended; they should be guided by the child's clinical condition and	GRADE D*	4	9 [5–9] APPROPRIATE (Relatively Strong Agreement)

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(*continued*)

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
	clinical scenario and/or previous imaging results and the ability to perform ultra-low dose free-breathing CT scans			

*RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

This statement is further supported by the following: There were no significant, meaningful changes in structural lung disease measured by chest CT at 1 and 3 years of age as assessed by PRAGMA-CF [70]. The median results at 1 and 3 years of age with CwCF showed a very low degree of bronchiectasis [70]. The extent of air trapping on CT (%TA) was low and was related to the extent of structural lung disease and LCI, but not to hyperinflation (increased FRC/TLC ratio measured on CT). There are also marked differences in the rate of progression, with subjects often showing improvement in lung disease [81].

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
10	In preschool children who are clinically unwell (with declining lung function and increased LCI, if available), a follow-up chest CT scan should be performed.	GRADE D*	4	9 [5–9] APPROPRIATE (Relatively Strong Agreement)

*RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
11	The time interval between 2 consecutive CT scans must be adapted to the clinical picture	GRADE D*	4	9 [8–9] APPROPRIATE (Strong Agreement)

*RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
12	School-age children with significant structural abnormalities should have repeat chest CT scans (rather than chest X-rays) every 2 or 3 years, even if they remain clinically healthy	GRADE D*	4	8 [1–9] UNCERTAIN (No consensus)

*RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

Other protocols are proposed in individual centres, depending on the possibility of performing ultra-low dose free-breathing CT scans and/or contrast MRIs of the lung to evaluate specific treatments [76,82]. In cooperative children with CF, they recommend performing an end-inspiratory and end-expiratory non-contrast-enhanced CT scan. Such teams should be able to use low-dose CT protocols and strategies that utilize similar radiation doses to those used in chest radiography to monitor patients with bronchiectasis [74].

5.2. *In adults*

De novo diagnoses of CF in adults account for up to 7 % of all new cases [83]. Pulmonary manifestations are rare in this population.

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
13	A *baseline* chest-CT should be performed at the time of a new diagnosis of CF in adults.	GRADE D*	4	9 [5–9] APPROPRIATE (Relatively Strong Agreement)

*RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

In patients with acute exacerbations, imaging should be performed in the clinical context, considering the cumulative radiation dose. Chest radiographs are less sensitive for selecting the underlying disease but are a useful tool for monitoring clinical response [6]. In patients with clinical progression where lung transplantation is a viable option, imaging should be in accordance with the healthcare system's pre-transplant protocol. Since chest radiographs are not sensitive enough to visualise changes in bronchial wall thickness or interstitial abnormalities, CT should be the preferred choice [84]. Technical progress should be considered while adhering to the ALARA (As Low as Reasonably Achievable) principle. A well-performed and dose-guided chest-CT every two years could reveal early signs of progressive disease. However, this was not retained during the Delphi process.

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
14	In adult patients without respiratory complications, chest CT imaging should be considered every 3–4 years.	GRADE D*	4	8 [1–9] UNCERTAIN (No consensus)
15	Imaging *follow-up* should be performed according to the clinical picture in adults.	GRADE D*	4	9 [5–9] APPROPRIATE (Relatively Strong Agreement)

*RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

Low-dose CTs may not be the optimal choice in adults. There is limited evidence on optimal techniques for this population. Options using photon-counting CT techniques are not yet widely available, but studies should be conducted to evaluate their use in clinical practice [85].

6. Harmonizing imaging protocols

6.1. *Rationale*

Most studies report improved detection of early lung disease, especially air trapping, in PwCF when expiratory volumetric settings are used in chest-CT. Expiratory volumetric settings may provide additional information, particularly in relation to air trapping, an early finding in PwCF that may allow for immediate diagnosis and treatment and potentially alter the course of disease [86,87]. However, the increased

radiation exposure associated with the addition of expiratory volumetric settings raises concerns about the risk-benefit ratio.

CT has been shown to be more sensitive than pulmonary function tests for detecting early and progressive lung disease in cooperative and uncooperative PwCF [88–90]. Chest-CT in CF includes both inspiratory and expiratory phases, which are recorded during periods of voluntary breath-holding. The end-inspiratory phase of chest-CT is primarily used to detect structural changes in the lungs, including bronchiectasis, peri-bronchial thickening, and consolidation. In contrast, expiratory volumetric settings aid in the detection of air trapping indicative of distal small airway disease, a potential early indicator of lung disease in CF [86,87]. This appears as hypodense areas reflecting regions of hyperinflation and/or hypoperfusion [87]. This results in the imaging sign seen on CT during air trapping, namely mosaic attenuation, which has been defined by the Fleischner Society as a "*patchwork of regions of varying attenuation in the lung*" and is characterised by heterogeneous lung attenuation with clearly defined borders corresponding to secondary lobules [91]. Along with bronchiectasis, air trapping has been associated with later progression of structural lung disease in a study of CwCF [70,92].

However, it remains uncertain whether areas of air trapping indicate a permanent obstruction of the small airways or a fluctuating and reversible condition [93]. Previous literature has emphasised that approximately one third of cases persist over time, suggesting irreversibility [94], however effective therapy has shown some degree of reversibility [62]. Nonetheless, CT imaging with expiratory volumetric settings is an effective modality for monitoring changes in air trapping before and after treatment [95]. However, it remains unclear whether CT is a more accurate tool for tracking small airway pathology compared to spirometry or parameters obtained by multiple gas washout [93]. LCI has been found to correlate with mosaic lung on CT, which indicates areas of air trapping/hypoperfusion [96,97]. Over the past decade, measurements of LCI and gas trapping have been increasingly favoured over FEV_1 for early monitoring of CF lung disease progression. This shift reflects their utility in tracking both radiological changes and clinical manifestations associated with disease progression [97–99]. The assessment of air trapping has been facilitated by several visual scoring systems over the past two decades, including the Bhalla [100], Brody II [101] and CF-CT [102].

6.2. Expiratory vs. inspiratory & expiratory volumetric chest-CT: clinical benefits/techniques/training modalities

The primary concern associated with recurrent CT imaging to monitor lung disease in CwCF is repeated exposure to ionising radiation. It is suggested that the radiation dose for expiratory CT scans should be set at 50 % less than for inspiratory CT scans [103]. In a cohort analysis of CwCF who underwent ultra-low-dose CT over the age of five years, a strong correlation between end-inspiratory and end-expiratory CT scores was observed, suggesting that end-expiratory alone may be sufficient, allowing a potential dose reduction of up to 75 % [104]. In a French study, 78 CT scans of 57 children aged 5–18 years were assessed using the full protocol images and expiratory sequence images only [105]. The correlations between the Brody global scores of the two different CT protocols were very good ($r = 0.90$ for both observers), for the bronchiectasis score ($r = 0.72$ and 0.86), the mucus plugging score ($r = 0.87$ and 0.83) and the expiratory air trapping ($r = 0.96$ and 0.92) [105]. The total dose of ionising radiation was reduced, with the measured dose-length product (DLP) reduced from 103.31 mGy cm (full protocol) to 3.06 mGy cm (expiratory protocol) ($p < 0.001$) [106]. It is therefore possible to complete inspiratory and expiratory chest CT scans to reveal early lung changes. In PwCF with more advanced changes, i.e. already developed bronchiectasis, inspiratory scans are considered more important than expiratory scans, and expiratory-only scans are not warranted.

It is noteworthy that the benefits of respiratory gating (a radiation therapy technique that uses computer software, a chest marker and infrared camera to track breathing) have mainly been demonstrated in children. In adults with CF the benefit of expiratory gating has not yet been clearly demonstrated [79].

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
16	The incorporation of expiratory views in longitudinal chest-CT follow-up assessments of PwCF should be considered.	Grade D*	4	7.5 [1–9] UNCERTAIN (No consensus)

*RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

6.3. Is sedation mandatory? What alternatives are there?

The radiologist decides whether sedation is necessary, taking into account the clinical context, the child's age and their ability to cooperate. The rapid performance of a CT scan allows it to be performed without anaesthesia in most cases, eliminating the complicated logistics and associated recovery times [6]. Therefore, it is often possible to perform free-breathing, non-enhanced CT [5,103], even in uncooperative children, with the newer fast CT-devices [76]. This is a key advantage over MRI, which requires moderate sedation for uncooperative patients [6].

In a survey conducted in 27 UK centres, 27 % reported that sedation was used because patients were uncooperative, previous CT without sedation failed or there was a learning disability. Reportedly 73 % used general anaesthesia in patients who were uncooperative and holding their breath, had a learning disability or were young [5].

6.4. Preparation of young children for CT

In cooperative school aged children and above, chest-CT should be regulated by spirometry to achieve a volume level close to the residual volume, in order to maximize the contrast between normal lung tissue and regions with low density [107]. However, institutions will require an adequately trained lung function technician/nurse to optimise results, which may not be logistically possible in all institutions [108]. Paediatric patients aged three to six years can be successfully taught breathing techniques by a trained technician in approximately half cases, without requiring spirometry guidance [108]. In uncooperative young paediatric patients, CT scans are typically performed with free breathing at their functional residual capacity (FRC). Typically, areas of air-trapping can be identified as the FRC level in these children is closer to the residual volume (RV) level compared to older children. Similarly, expiratory scans in anesthetized children are also performed at the FRC level, given that there is no active expiration to the RV level [108].

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
17	In cooperative children of school age and older, lung volume control during chest-CT should be optimised through coaching by staff trained in this technique.	Grade C*	2+	8.5 [5–9] APPROPRIATE (Relatively Strong Agreement)
18	For research purposes,	Grade D**	4	9 [5–9] APPROPRIATE

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Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
	ideally a technician/ staff member trained in volume optimization should be present to supervise the subject and check that they are maintaining an optimal respiratory rate (with or without spirometry) during the CT scan.			(Relatively Strong Agreement)

*RECOMMENDATION OF MODERATE CERTAINTY.
**RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

7. Reporting of results

The severity of lung disease can be assessed either subjectively, based on the radiologist's experience, or more objectively, using visual or automated scoring methods. Several visual scoring methods have been proposed to quantify CF-related morphological abnormalities in CT imaging. These methods are based on quantifying the extent and severity of the main morphological abnormalities, which include bronchiectasis, bronchial wall thickening, mucoid impaction and mosaic perfusion/air trapping. The three main visual scoring methods are the Bhalla score [100,109,110], the Brody score (derived from the score proposed by Mafessanti et al.) and its derivatives [55,109,111,112] and the PRAGMA-CF score [64,65,66,70,113].

The use of scoring methods in clinical practice implies a benefit at the individual level. Their intra- and inter-observer reproducibility is excellent, but requires specific training, and reproducibility decreases over time [114]. They can be performed in 10–15 min [20], which is time consuming but potentially acceptable, and they can discriminate multiple severity levels [21]. However, the impact of visual scores on the management of PwCF patients has not yet been demonstrated, and their ability to detect longitudinal changes in disease severity at the individual level is uncertain. The role of visual CT scores in clinical practice, particularly in patient monitoring, therefore, remains to be established.

Automated scoring methods are a promising alternative to visual CT scores [115,116]. The advantage of these automated CT scores is that they can be calculated within seconds to minutes with perfect reproducibility. However, for most of these scores, the sensitivity to longitudinal change in disease severity has not been evaluated. Visual chest CT scores can be used to assess disease severity, but there is insufficient evidence to recommend any particular scoring system. Automated scoring methods require longitudinal validation. Current visual scores are unsuitable for assessing improvement on an individual scale.

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
19	Current visual scores (e.g. Bhalla, Brody, PRAGMA-CF, etc.) can be used to assess disease	GRADE D*	4	8 [5–9] APPROPRIATE (Relatively Strong Agreement)

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(*continued*)

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
	severity, but are less useful for assessing longitudinal improvement at an individual level, as their impact on patient management has not yet been demonstrated.			

*RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

The radiology report can be free text or structured. Whilst free text is prone to heterogeneity, structured reports aim to standardise organization and language, which may improve reporting of relevant findings and communication with referring physicians [117–120]. However, a clear benefit of its use over the free text has not yet been demonstrated in clinical practice. Although pulmonologists and radiologists favoured adopting structured reporting, less than 20 % used it in practice. There is still insufficient data to support its systematic implementation. Further studies could explore variability by expertise and assess its added value for longitudinal evaluation of disease.

Statement n°	Statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
20	A structured chest CT report in CF is currently not generally recommended as further studies are still pending	GRADE D*	4	7 [1–9] UNCERTAIN (No Consensus)

*RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

8. 'Added value' of CT-scans and morphomics

Clinical teams should consider the added value that imaging can provide beyond its original purpose. For example, CT chest scans whose clinical indication is to assess the respiratory system may yield additional information about body composition, cardiovascular risk or bone health, which will be of increasing value in a complex, ageing CF population [121]. In a cross-sectional study, CT measurements of muscle mass at T4 and T12 in adults showed a significant correlation with handgrip strength and fat-free mass index. T12 also showed a significant correlation with FEV_1 and not with body mass index [121]. In another longitudinal study of adult PwCF on Elexacaftor/Tezacaftor/Ivacaftor, in addition to a significant increase in BMI, body composition also changed with an increase in the cross-sectional area of subcutaneous, thoracic and visceral upper abdominal fat. Even in those who had a normal BMI at the time of follow-up, epicardial and upper abdominal visceral fat and subcutaneous fat had increased significantly over the interval [S. Blackburn 2024, personal data].

9. The future

Improvements in therapeutic management (modulator and gene therapies) and in the performance of CT-devices, allowing for minimal radiation exposure, are to be expected in the future.

The advent of CFTR modulator therapy (MT) will change the long-term course of CF lung disease [122]. Already, clinicians caring for PwCF recognise different groups within the modulator-available population: children and adolescents who started MT before the onset of

severe irreversible lung damage, young adults with improved moderate lung changes and adults with advanced CF lung disease at the onset of MT in whom stabilisation has been achieved. However, the long-term effects of MT are currently unclear, so the frequency of imaging must be weighed against the radiation risk.

When it comes to imaging, there is ongoing research between newer chest CT methods and radiation-free MRI. An example of the former is photon counting CT. In recent years, CT technology has evolved with the newly available photon counting detectors, representing further progress in both radiation reduction and spatial resolution. In contrast to conventional energy integrating detectors (EID), which use scintillator detectors to convert photons into light signals, photon counting detectors convert photon energy directly into electrical signals using cadmium electrode semiconductors [123,124]. Photon-counting CTs (PCCTs) have now become routine clinical practice in a few specialised centres, with initial studies showing that LD-HR PCCT lung scans provide comparable or better image quality while requiring significantly lower radiation doses compared to EID CT scans [125–128]. Dettmer et al. demonstrated that it is possible to achieve the radiation dose corresponding to a CXR as defined in the German diagnostic reference value for CXR of 52 cGy cm^2 (12 cGy cm^2 for a posterior-anterior and 40 cGy cm^2 for a lateral exposure) with a ULD thoracic protocol on the PCCT [129,130]. This suggests that CT will replace CXR in certain clinical scenarios due to its improved diagnostic capabilities. In addition, due to its low radiation exposure, photon counting CT could serve as a baseline and follow-up examination in paediatrics, and emergency situations, as well as when MRI availability is limited [131].

10. Conclusion

We recommend the use of ultra-low dose CT scans and a system for tracking cumulative radiation per patient. Our guidance defines effective radiation doses and radiation risk in order to further fine-tune radiological techniques in European CF centres. Even if individual radiation risks are quite low, radiation protection in imaging is a public health issue due to the large population exposed to these risks. Children can be two to three times more sensitive to radiation than adults [132]. When indicated and available, imaging techniques without ionising radiation, e.g. ultrasound (sound waves) or MRI (radiofrequency and electromagnetic waves), should be preferred, especially in very young preschool children and pregnant women.

Potential limitations of this guidance include the variability in consensus in the Delphi process and the low to moderate level of evidence for many of the recommendations. These limitations highlight the challenges in this evolving field of study and reinforce the need for further research in this area, such as the importance of automated techniques to facilitate the search for minimally clinically meaningful differences for relevant outcomes and disease trajectories particularly in the era of CF modulator therapy. Moreover, this guidance stresses the importance of being more user-friendly to reduce the burden of imaging and radiation risk. Patients and families should be involved in the risk–benefit discussion about imaging procedures so that they can best understand the information and make an informed decision [132].

Ultimately, indications will depend on the ability of radiologists and physicists in the field to perform chest and other CT scans at ultra-low doses and to track lifetime risk using radiation passports. The final decision may also be influenced by cost, expertise, availability of resources and/or the patient's values, as well as a possible change in treatment.

CRediT authorship contribution statement

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Data availability statement

No patient data associated with this guidance document. Any information associated with the Delphi process was anonymised and is confidential.

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References

[1] C. Crowley, O.J.O. Connor, P. Ciet, H. Tiddens, M.M. Maher, The evolving role of radiological imaging in cystic fibrosis, Curr. Opin. Pulm. Med. 27 (6) (2021) 575–585.

[2] H. Ferris, M. Twomey, F. Moloney, S.B. O'Neill, K. Murphy, O.J. O'Connor, et al., Computed tomography dose optimisation in cystic fibrosis: a review, World J. Radiol. 8 (4) (2016) 331–341.

[3] M. Stahl, Sensitive markers to detect progression of lung disease in children with cystic fibrosis, Eur. Respir. J. 58 (1) (2021).

[4] K.J. Bayfield, T.A. Douglas, T. Rosenow, J.C. Davies, S.J. Elborn, M. Mall, et al., Time to get serious about the detection and monitoring of early lung disease in cystic fibrosis, Thorax 76 (12) (2021) 1255–1265.

[5] F.J. Gilchrist, R. Buka, M. Jones, S.A. Ho, W. Lenney, W.D. Carroll, Clinical indications and scanning protocols for chest CT in children with cystic fibrosis: a survey of UK tertiary centres, BMJ Paediatr Open 2 (1) (2018) e000367.

[6] P. Ciet, S. Bertolo, M. Ros, R. Casciaro, M. Cipolli, S. Colagrande, et al., State-of-the-art review of lung imaging in cystic fibrosis with recommendations for pulmonologists and radiologists from the "iMAging managEment of cySTic fibROsis" (MAESTRO) consortium, Eur. Respir. Rev. 31 (163) (2022).

[7] L. Bastiani, F. Paolicchi, L. Faggioni, M. Martinelli, R. Gerasia, C. Martini, et al., Patient perceptions and knowledge of ionizing radiation from medical imaging, JAMA Netw. Open 4 (10) (2021) e2128561.

[8] R. Rai, S. Kumar, V. Batumalai, D. Elwadia, L. Ohanessian, E. Juresic, et al., The integration of MRI in radiation therapy: collaboration of radiographers and radiation therapists, J. Med. Radiat Sci. 64 (1) (2017) 61–68.

[9] S.D. Brown, M.J. Callahan, Beyond the AJR: enduring gaps in patient understanding of imaging-associated ionizing radiation, AJR Am. J. Roentgenol. 219 (1) (2022) 170.

[10] F.T. Delaney, T.O. Doinn, J.M. Broderick, E. Stanley, Readability of patient education materials related to radiation safety: what are the implications for patient-centred radiology care? Insights Imag. 12 (1) (2021) 148.

[11] A.F. Reitan, A. Sanderud, Communicating radiation risk to patients: experiences among radiographers in Norway, J. Med. Imag. Radiat. Sci. 51 (4S) (2020) S84–S89.

[12] L. Ukkola, H. Kyngas, A. Henner, H. Oikarinen, Barriers to not informing patients about radiation in connection with radiological examinations: radiographers' opinion, Radiography 26 (2) (2020) e114–e119.

[13] E. Kenny, B. Byrne, M. Lewis, D.M. King, Perception of medical radiation risk in Ireland: results of a public survey, Phys. Med. 68 (2019) 96–103.

[14] C. Stewart, R. Smith-Bindman, It is time to inform patients of medical imaging risks, JAMA Netw. Open 4 (10) (2021) e2129681.

[15] R.S.N.A. Acr, A.A.P.M. Asrt, Image wisely, Available from: https://www.imagewisely.org/Imaging-Modalities/Computed-Tomography, 2023.

[16] V.J. Earl, A.O.G. Potter, A.A. Perdomo, Effective doses for common paediatric diagnostic general radiography examinations at a major Australian paediatric hospital and the communication of associated radiation risks, J. Med. Radiat Sci. 70 (1) (2023) 30–39.

[17] D.J. Brenner, E.J. Hall, Computed tomography — an increasing source of radiation exposure, N. Engl. J. Med. 357 (22) (2007) 2277–2284.

[18] M.F. McNitt-Gray, AAPM/RSNA physics tutorial for residents: topics in CT: radiation dose in CT, Radiographics 22 (6) (2002) 1541–1553.

[19] P.A. De Jong, H.A.W.M. Tiddens, Cystic fibrosis specific computed tomography scoring, Proc. Am. Thorac. Soc. 4 (4) (2007) 338–342.

[20] N. Begum, C.A. Byrnes, J. Cheney, P.J. Cooper, E. Fantino, N. Gailer, et al., Factors in childhood associated with lung function decline to adolescence in cystic fibrosis, J. Cyst. Fibros. 21 (6) (2022) 977–983.

[21] S. Joyce, B.W. Carey, N. Moore, D. Mullane, M. Moore, M.F. McEntee, et al., Computed tomography in cystic fibrosis lung disease: a focus on radiation exposure, Pediatr. Radiol. 51 (4) (2021) 544–553.

[22] F. Moloney, R.G. Kavanagh, N.J. Ronan, T.M. Grey, S. Joyce, D.J. Ryan, et al., 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 (5) (2021) 393, e9- e17.

[23] A.A. Bankier, H.Y. Kressel, Through the looking glass revisited: the need for more meaning and less drama in the reporting of dose and dose reduction in CT, Radiology 265 (1) (2012) 4–8.

[24] O.J. O'Connell, S. McWilliams, A. McGarrigle, O.J. O'Connor, F. Shanahan, D. Mullane, et al., Radiologic imaging in cystic fibrosis, Chest 141 (6) (2012) 1575–1583.

[25] M. Ponnusamy, D. Halanaik, V. Rajaraman, Size specific dose estimate (SSDE) for estimating patient dose from CT used in myocardial perfusion SPECT/CT, Asia Oceania J. Nuclear Med. Biolog. (2019) (Online First).

[26] C.H. McCollough, L. Guimarães, J.G. Fletcher, In defense of body CT, Am. J. Roentgenol. 193 (1) (2009) 28–39.

[27] P. Mohammadinejad, A. Mileto, L. Yu, S. Leng, L.S. Guimaraes, A.D. Missert, et al., CT noise-reduction methods for lower-dose scanning: strengths and weaknesses of iterative reconstruction algorithms and new techniques, Radiographics 41 (5) (2021) 1493–1508.

[28] F.R. Verdun, D. Gutierrez, P. Schnyder, A. Aroua, F. Bochud, F. Gudinchet, CT dose optimization when changing to CT multi-detector row technology, Curr. Probl. Diagn. Radiol. 36 (4) (2007) 176–184.

[29] C.H. Clement, International commission on radiological protection: adult reference computational phantoms, Available from: https://www.icrp.org/publication.asp?id=icrp%20publication%20110, 2009.

[30] International Commission on Radiological Protection (ICRP), Paediatric Reference Computational Phantoms, ICRP Publication, 2020, p. 49.

[31] The measurement, reporting, and management of radiation dose in CT, Report of AAPM Task Group 23, Report 96 (2008) 1–34. https://www.aapm.org/pubs/reports/rpt_96.pdf.

[32] R. Smith-Bindman, Radiation dose associated with common computed tomography examinations and the associated lifetime attributable risk of cancer, Arch. Intern. Med. 169 (22) (2009) 2078.

[33] P.D. Deak, Y. Smal, W.A. Kalender, Multisection CT protocols: sex- and age-specific conversion factors used to determine effective dose from dose-length product, Radiology 257 (1) (2010) 158–166.

[34] N.J. Ronan, G.G. Einarsson, M. Twomey, D. Mooney, D. Mullane, M. NiChroinin, et al., CORK study in cystic fibrosis, Chest 153 (2) (2018) 395–403.

[35] F. Homayounieh, O. Holmberg, R.A. Umairi, S. Aly, A. Basevicius, P.R. Costa, et al., Variations in CT utilization, protocols, and radiation doses in COVID-19 pneumonia: results from 28 countries in the IAEA study, Radiology 298 (3) (2021) E141–E151.

[36] J.E. Lee, S.Y. Choi, J.A. Hwang, S. Lim, M.H. Lee, B.H. Yi, et al., The potential for reduced radiation dose from deep learning-based CT image reconstruction: a comparison with filtered back projection and hybrid iterative reconstruction using a phantom, Medicine (Baltim.) 100 (19) (2021) e25814.

[37] W. Huda, CT dose metrics, Radiology 267 (3) (2013) 964–967.

[38] F.R. Verdun, F. Bochud, F. Gundinchet, A. Aroua, P. Schnyder, R. Meuli, Quality initiatives radiation risk: what you should know to tell your patient, Radiographics 28 (7) (2008) 1807–1816.

[39] S.J. Westra, The communication of the radiation risk from CT in relation to its clinical benefit in the era of personalized medicine: Part 2: benefits versus risk of CT, Pediatr. Radiol. 44 (S3) (2014) 525–533.

[40] E.C. Lin, Radiation risk from medical imaging, Mayo Clin. Proc. 85 (12) (2010) 1142–1146.

[41] A. Martinez Lorca, R. Aguado Romo, M. Martinez Lorca, M.C. Zabala Banos, Anxiety reduction and emotional self-care using the U-technique in radiology departments, Br. J. Radiol. 90 (1078) (2017) 20170173.

[42] L. Vieira, A. Pires, A. Grilo, Anxiety experienced by oncological patients who undergo 18F-FDG PET CT: a systematic review, Radiography 27 (4) (2021) 1203–1210.
[43] J.E. Middelkamp, B.B. Forster, C. Keogh, P. Lennox, K. Mayson, Evaluation of adult outpatient magnetic resonance imaging sedation practices: are patients being sedated optimally? Can. Assoc. Radiol. J. 60 (4) (2009) 190–195.
[44] A. Martinez Lorca, R. Aguado Romo, M. Martinez Lorca, M.C. Zabala Baños, Anxiety reduction and emotional self-care using the U-technique in radiology departments, Br. J. Radiol. 90 (1078) (2017) 20170173.
[45] J.E.M. Madl, S.C. Sturmbauer, R. Janka, S. Bay, N. Rohleder, Preparing patients according to their individual coping style improves patient experience of magnetic resonance imaging, J. Behav. Med. 45 (6) (2022) 841–854.
[46] D.D. Walworth, Effect of live music therapy for patients undergoing magnetic resonance imaging, J. Music Ther. 47 (4) (2010) 335–350.
[47] K. Shimokawa, K. Matsumoto, H. Yokota, E. Kobayashi, Y. Hirano, Y. Masuda, et al., Anxiety relaxation during MRI with a patient-friendly audiovisual system, Radiography 28 (3) (2022) 725–731.
[48] J. Vassileva, O. Holmberg, Radiation protection perspective to recurrent medical imaging: what is known and what more is needed? Brit J Radiol 94 (1126) (2021) 20210477.
[49] R. Seuri, M.M. Rehani, M. Kortesniemi, How tracking radiologic procedures and dose helps: experience from Finland, Am. J. Roentgenol. 200 (4) (2013) 771–775.
[50] M. Brambilla, J. Vassileva, A. Kuchcinska, M.M. Rehani, Multinational data on cumulative radiation exposure of patients from recurrent radiological procedures: call for action, Eur. Radiol. 30 (5) (2020) 2493–2501.
[51] O. Holmberg, J. Malone, M. Rehani, D. McLean, R. Czarwinski, Current issues and actions in radiation protection of patients, Eur. J. Radiol. 76 (1) (2010) 15–19.
[52] M.M. Rehani, Patient radiation exposure and dose tracking: a perspective, J. Med. Imaging 4 (3) (2017) 031206.
[53] M.O. Baerlocher, R. Talanow, A.F. Baerlocher, Radiation passport: an iPhone and iPod touch application to track radiation dose and estimate associated cancer risks, J. Am. Coll. Radiol. 7 (4) (2010) 277–280.
[54] X. Sun, H. Sui, J.T. Fisher, Z. Yan, X. Liu, H.J. Cho, et al., Disease phenotype of a ferret CFTR-knockout model of cystic fibrosis, J. Clin. Investig. 120 (9) (2010) 3149–3160.
[55] P.A. de Jong, A. Lindblad, L. Rubin, W.C. Hop, J.C. de Jongste, M. Brink, et al., Progression of lung disease on computed tomography and pulmonary function tests in children and adults with cystic fibrosis, Thorax 61 (1) (2006) 80–85.
[56] P.D. Sly, S. Brennan, C. Gangell, N. de Klerk, C. Murray, L. Mott, et al., Lung disease at diagnosis in infants with cystic fibrosis detected by newborn screening, Am. J. Respir. Crit. Care Med. 180 (2) (2009) 146–152.
[57] P.D. Sly, C.L. Gangell, L. Chen, R.S. Ware, S. Ranganathan, L.S. Mott, et al., Risk factors for bronchiectasis in children with cystic fibrosis, N. Engl. J. Med. 368 (21) (2013) 1963–1970.
[58] M.O. Wielputz, M. Puderbach, A. Kopp-Schneider, M. Stahl, E. Fritzsching, O. Sommerburg, et al., 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 (8) (2014) 956–965.
[59] S.D. Davis, L.A. Fordham, A.S. Brody, T.L. Noah, G.Z. Retsch-Bogart, B.F. Qaqish, et al., Computed tomography reflects lower airway inflammation and tracks changes in early cystic fibrosis, Am. J. Respir. Crit. Care Med. 175 (9) (2007) 943–950.
[60] M. Stahl, E. Steinke, S.Y. Graeber, C. Joachim, C. Seitz, H.U. Kauczor, et al., Magnetic resonance imaging detects progression of lung disease and impact of newborn screening in preschool children with cystic fibrosis, Am. J. Respir. Crit. Care Med. 204 (8) (2021) 943–953.
[61] S.C. Ranganathan, G.L. Hall, P.D. Sly, S.M. Stick, T.A. Douglas, Australian respiratory early surveillance team for cystic F. Early lung disease in infants and preschool children with cystic fibrosis. What have we learned and what should we do about it? Am. J. Respir. Crit. Care Med. 195 (12) (2017) 1567–1575.
[62] T.E. Robinson, M.L. Goris, H.J. Zhu, X. Chen, P. Bhise, F. Sheikh, et al., Dornase alfa reduces air trapping in children with mild cystic fibrosis lung disease, Chest 128 (4) (2005) 2327–2335.
[63] F. Ratjen, S.D. Davis, S. Stanojevic, R.A. Kronmal, K.D. Hinckley Stukovsky, N. Jorgensen, et al., Inhaled hypertonic saline in preschool children with cystic fibrosis (SHIP): a multicentre, randomised, double-blind, placebo-controlled trial, Lancet Respir. Med. 7 (9) (2019) 802–809.
[64] H. Tiddens, Y. Chen, E.R. Andrinopoulou, S.D. Davis, M. Rosenfeld, F. Ratjen, et al., The effect of inhaled hypertonic saline on lung structure in children aged 3-6 years with cystic fibrosis (SHIP-CT): a multicentre, randomised, double-blind, controlled trial, Lancet Respir. Med. 10 (7) (2022) 669–678.
[65] S.M. Stick, A. Foti, R.S. Ware, H. Tiddens, B.S. Clements, D.S. Armstrong, et al., The effect of azithromycin on structural lung disease in infants with cystic fibrosis (COMBAT CF): a phase 3, randomised, double-blind, placebo-controlled clinical trial, Lancet Respir. Med. 10 (8) (2022) 776–784.
[66] L.C. Mok, A. Garcia-Uceda, M.N. Cooper, M. Kemner-Van De Corput, M. De Bruijne, N. Feyaerts, et al., The effect of CFTR modulators on structural lung disease in cystic fibrosis, Front. Pharmacol. 14 (2023) 1147348.
[67] K. Newbegin, K. Pilkington, S. Shanthikumar, S. Ranganathan, Clinical utility of surveillance computed tomography scans in infants with cystic fibrosis, Pediatr. Pulmonol. 53 (10) (2018) 1387–1390.
[68] L.P. Thia, A. Calder, J. Stocks, A. Bush, C.M. Owens, C. Wallis, et al., Is chest CT useful in newborn screened infants with cystic fibrosis at 1 year of age? Thorax 69 (4) (2014) 320–327.
[69] G. Davies, L.P. Thia, J. Stocks, A. Bush, A.F. Hoo, A. Wade, et al., 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 (6) (2020) 896–901.
[70] T. Rosenow, M.C. Oudraad, C.P. Murray, L. Turkovic, W. Kuo, M. de Bruijne, et al., PRAGMA-CF. A quantitative structural lung disease computed tomography outcome in young children with cystic fibrosis, Am. J. Respir. Crit. Care Med. 191 (10) (2015) 1158–1165.
[71] R.M. Sandvik, T. Kongstad, K. Green, C. Voldby, F. Buchvald, M. Skov, et al., Prospective longitudinal association between repeated multiple breath washout measurements and computed tomography scores in children with cystic fibrosis, J. Cyst. Fibros. 20 (4) (2021) 632–640.
[72] D. Caudri, L. Turkovic, N.H. de Klerk, T. Rosenow, C.P. Murray, E.W. Steyerberg, et al., A screening tool to identify risk for bronchiectasis progression in children with cystic fibrosis, Pediatr. Pulmonol. 57 (1) (2022) 122–131.
[73] W.H. Wang, C.Y. Sung, S.C. Wang, Y.J. Shao, Risks of leukemia, intracranial tumours and lymphomas in childhood and early adulthood after pediatric radiation exposure from computed tomography, CMAJ (Can. Med. Assoc. J.) 195 (16) (2023) E575–E583.
[74] K.P. Murphy, M.M. Maher, O.J. O'Connor, Imaging of cystic fibrosis and pediatric bronchiectasis, AJR Am. J. Roentgenol. 206 (3) (2016) 448–454.
[75] A. de Martini, R. Price, S. Romeo, D3.6 Report on patients' research needs and expectations in medical application of ionising radiation and radiation protection, EURAMED Rocc-N-Roll, aspects EMaaRpCsraaRithad (2022 NFRP-2019-2020).
[76] P. Ciet, R. Booij, M. Dijkshoorn, M. van Straten, H. Tiddens, Chest radiography and computed tomography imaging in cystic fibrosis: current challenges and new perspectives, Pediatr. Radiol. 53 (4) (2023) 649–659.
[77] A.R. Smyth, S.C. Bell, S. Bojcin, M. Bryon, A. Duff, P. Flume, et al., European cystic fibrosis society standards of care: best practice guidelines, J. Cyst. Fibros. 13 (Suppl 1) (2014) S23–S42.
[78] K.W. Southern, C. Addy, S. Bell, A. Bevan, U. Borawska, C. Brown, et al., Standards for the care of people with cystic fibrosis; establishing and maintaining health, J. Cyst. Fibros. 23 (1) (2024) 12–28.
[79] E. Salamon, S. Lever, W. Kuo, P. Ciet, H.A. Tiddens, Spirometer guided chest imaging in children: it is worth the effort, Pediatr. Pulmonol. 52 (1) (2017) 48–56.
[80] L.A. Tepper, D. Caudri, A.P. Rovira, H.A. Tiddens, M. de Bruijne, The development of bronchiectasis on chest computed tomography in children with cystic fibrosis: can pre-stages be identified? Eur. Radiol. 26 (12) (2016) 4563–4569.
[81] A.S. Brody, R. Huang, B. Zhang, F.R. Long, S.W. Powers, Structural lung disease in preschool children with cystic fibrosis: an 18 month natural history study, J. Cyst. Fibros. 21 (2) (2022) e165–e171.
[82] N. Landini, P. Ciet, H.M. Janssens, S. Bertolo, M. Ros, M. Mattone, et al., Management of respiratory tract exacerbations in people with cystic fibrosis: focus on imaging, Front Pediatr 10 (2022) 1084313.
[83] S. Averill, M.G. Lubner, C.O. Menias, S. Bhalla, V.M. Mellnick, T.A. Kennedy, et al., Multisystem imaging findings of cystic fibrosis in adults: recognizing typical and atypical patterns of disease, AJR Am. J. Roentgenol. 209 (1) (2017) 3–18.
[84] W. Kuo, P. Ciet, H.A. Tiddens, W. Zhang, R.P. Guillerman, M. van Straten, Monitoring cystic fibrosis lung disease by computed tomography. Radiation risk in perspective, Am. J. Respir. Crit. Care Med. 189 (11) (2014) 1328–1336.
[85] K.K. Horst, N.C. Hull, P.G. Thacker, N. Demirel, L. Yu, J.S. McDonald, et al., Pilot study to determine whether reduced-dose photon-counting detector chest computed tomography can reliably display Brody II score imaging findings for children with cystic fibrosis at radiation doses that approximate radiographs, Pediatr. Radiol. 53 (6) (2023) 1049–1056.
[86] T.E. Robinson, M.L. Goris, R.B. Moss, L. Tian, P. Kan, M. Yilma, et al., Mucus plugging, air trapping, and bronchiectasis are important outcome measures in assessing progressive childhood cystic fibrosis lung disease, Pediatr. Pulmonol. 55 (4) (2020) 929–938.
[87] H.A.W.M. Tiddens, S.M. Stick, S. Davis, Multi-modality monitoring of cystic fibrosis lung disease: the role of chest computed tomography, Paediatr. Respir. Rev. 15 (1) (2014) 92–97.
[88] L. Turkovic, D. Caudri, T. Rosenow, O. Breuer, C. Murray, H. Tiddens, et al., Structural determinants of long-term functional outcomes in young children with cystic fibrosis, Eur. Respir. J. 55 (5) (2020).
[89] N.E. Wijker, S. Vidmar, K. Grimwood, P.D. Sly, C.A. Byrnes, J.B. Carlin, et al., Early markers of cystic fibrosis structural lung disease: follow-up of the ACFBAL cohort, Eur. Respir. J. 55 (4) (2020).
[90] P.A. de Jong, Y. Nakano, M.H. Lequin, J.R. Mayo, R. Woods, P.D. Pare, et al., Progressive damage on high resolution computed tomography despite stable lung function in cystic fibrosis, Eur. Respir. J. 23 (1) (2004) 93–97.
[91] J.M. Bryant, D.M. Grogono, D. Greaves, J. Foweraker, I. Roddick, T. Inns, et al., Whole-genome sequencing to identify transmission of Mycobacterium abscessus between patients with cystic fibrosis: a retrospective cohort study, Lancet 381 (9877) (2013) 1551–1560.
[92] N. Pillarisetti, B. Linnane, S. Ranganathan, Early bronchiectasis in cystic fibrosis detected by surveillance CT: case Reports, Respirology 15 (6) (2010) 1009–1011.
[93] H.A.W.M. Tiddens, S.H. Donaldson, M. Rosenfeld, P.D. Paré, Cystic fibrosis lung disease starts in the small airways: can we treat it more effectively? Pediatr. Pulmonol. 45 (2) (2010) 107–117.
[94] M. Loeve, T. Rosenow, V. Gorbunova, W.C. Hop, H.A. Tiddens, M. de Bruijne, Reversibility of trapped air on chest computed tomography in cystic fibrosis patients, Eur. J. Radiol. 84 (6) (2015) 1184–1190.
[95] R. Bec, M. Reynaud-Gaubert, F. Arnaud, R. Naud, N. Dufeu, M. Di Bisceglie, et al., Chest computed tomography improvement in patients with cystic fibrosis treated

with elexacaftor-tezacaftor-ivacaftor: early report, Eur. J. Radiol. 154 (2022) 110421.
[96] S. Verbanck, G.G. King, W. Zhou, A. Miller, C. Thamrin, D. Schuermans, et al., The quantitative link of lung clearance index to bronchial segments affected by bronchiectasis, Thorax 73 (1) (2018) 82–84.
[97] T. Rosenow, K. Ramsey, L. Turkovic, C.P. Murray, L.C. Mok, G.L. Hall, et al., Air trapping in early cystic fibrosis lung disease-Does CT tell the full story? Pediatr. Pulmonol. 52 (9) (2017) 1150–1156.
[98] K.A. Ramsey, C. McGirr, S.M. Stick, G.L. Hall, S.J. Simpson, Effect of posture on lung ventilation distribution and associations with structure in children with cystic fibrosis, J. Cyst. Fibros. 16 (6) (2017) 713–718.
[99] S.A. Rowan, J.M. Bradley, I. Bradbury, J. Lawson, T. Lynch, P. Gustafsson, et al., Lung clearance index is a repeatable and sensitive indicator of radiological changes in bronchiectasis, Am. J. Respir. Crit. Care Med. 189 (5) (2014) 586–592.
[100] M. Bhalla, N. Turcios, V. Aponte, M. Jenkins, B.S. Leitman, D.I. McCauley, et al., Cystic fibrosis: scoring system with thin-section CT, Radiology 179 (3) (1991) 783–788.
[101] A.S. Brody, H.A.W.M. Tiddens, R.G. Castile, H.O. Coxson, P.A. de Jong, J. Goldin, et al., Computed tomography in the evaluation of cystic fibrosis lung disease, Am. J. Respir. Crit. Care Med. 172 (10) (2005) 1246–1252.
[102] W. Kuo, E.R. Andrinopoulou, A. Perez-Rovira, H. Ozturk, M. de Bruijne, H. A. Tiddens, Objective airway artery dimensions compared to CT scoring methods assessing structural cystic fibrosis lung disease, J. Cyst. Fibros. 16 (1) (2017) 116–123.
[103] W. Kuo, M.P.C. Kemner-van de Corput, A. Perez-Rovira, M. de Bruijne, I. Fajac, H. A.W.M. Tiddens, et al., Multicentre chest computed tomography standardisation in children and adolescents with cystic fibrosis: the way forward, Eur. Respir. J. 47 (6) (2016) 1706–1717.
[104] M. Loeve, M.H. Lequin, M. De Bruijne, I.J.C. Hartmann, K. Gerbrands, M. Van Straten, et al., Cystic fibrosis: are volumetric ultra-low-dose expiratory CT scans sufficient for monitoring related lung disease? Radiology 253 (1) (2009) 223–229.
[105] S. Simon, C. Baunin, J. Vial, E. Mas, L. Roditis, M. Michelet, et al., [Computed tomography in children with cystic fibrosis: the role of an expiratory protocol], Rev. Mal. Respir. 37 (5) (2020) 355–363.
[106] J.C. Davies, C.E. Wainwright, G.S. Sawicki, M.N. Higgins, D. Campbell, C. Harris, et al., Ivacaftor in infants aged 4 to <12 Months with cystic fibrosis and a gating mutation. Results of a two-Part Phase 3 clinical trial, Am. J. Respir. Crit. Care Med. 203 (5) (2021) 585–593.
[107] T.E. Robinson, A.N. Leung, R.B. Moss, F.G. Blankenberg, H. al-Dabbagh, W. H. Northway, Standardized high-resolution CT of the lung using a spirometer-triggered electron beam CT scanner, Am. J. Roentgenol. 172 (6) (1999) 1636–1638.
[108] M. van Straten, A.S. Brody, C. Ernst, R.P. Guillerman, H.A.W.M. Tiddens, S. K. Nagle, Guidance for computed tomography (CT) imaging of the lungs for patients with cystic fibrosis (CF) in research studies, J. Cyst. Fibros. 19 (2) (2020) 176–183.
[109] T.H. Helbich, G. Heinz-Peer, I. Eichler, P. Wunderbaldinger, M. Gotz, C. Wojnarowski, et al., Cystic fibrosis: CT assessment of lung involvement in children and adults, Radiology 213 (2) (1999) 537–544.
[110] L. Dorlochter, H. Nes, G. Fluge, K. Rosendahl, High resolution CT in cystic fibrosis–the contribution of expiratory scans, Eur. J. Radiol. 47 (3) (2003) 193–198.
[111] A.S. Brody, Early morphologic changes in the lungs of asymptomatic infants and young children with cystic fibrosis, J. Pediatr. 144 (2) (2004) 145–146.
[112] M. Maffessanti, M. Candusso, F. Brizzi, F. Piovesana, Cystic fibrosis in children: HRCT findings and distribution of disease, J. Thorac. Imag. 11 (1) (1996) 27–38.
[113] P. McNally, K. Lester, G. Stone, B. Elnazir, M. Williamson, D. Cox, B. Linnane, L. Kirwan, D. Rea, P. O'Regan, T. Semple, C. Saunders, H.A.W.M. Tiddens, E. McKone, J.C. Davies, RECOVER Study Group, Improvement in lung clearance index and chest computed tomography scores with elexacaftor/tezacaftor/ivacaftor treatment in people with cystic fibrosis aged 12 Years and older - the RECOVER trial, Am. J. Respir. Crit. Care Med. 208 (9) (2023 Nov 1) 917–929, https://doi.org/10.1164/rccm.202308-1317OC. PMID: 37703083.
[114] P.A. de Jong, M.D. Ottink, S.G.F. Robben, M.H. Lequin, W.C.J. Hop, J.J. E. Hendriks, et al., Pulmonary disease assessment in cystic fibrosis: comparison of CT scoring systems and value of bronchial and arterial dimension measurements, Radiology 231 (2) (2004) 434–439.
[115] G. Dournes, C.S. Hall, M.M. Willmering, A.S. Brody, J. Macey, S. Bui, B. Denis de Senneville, P. Berger, F. Laurent, I. Benlala, J.C. Woods, Artificial intelligence in computed tomography for quantifying lung changes in the era of CFTR modulators, Eur. Respir. J. 59 (3) (2022 Mar 3) 2100844, https://doi.org/10.1183/13993003.00844-2021. PMID: 34266943.
[116] Y. Chen, J.-P. Charbonnier, E.-R. Andrinopoulou, P.D. Sly, S.M. Stick, H.A.W. M. Tiddens, on behalf of the COMBAT-CF Study Group, Azithromycin reduces bronchial wall thickening in infants with CF, J. Cyst. Fibros. 23 (2024) 870–873.
[117] European Society of Radiology (ESR), ESR paper on structured reporting in radiology, Insights into Imag. 9 (1) (2018) 1–7.
[118] D.B. Larson, A.J. Towbin, R.M. Pryor, L.F. Donnelly, Improving consistency in radiology reporting through the use of department-wide standardized structured reporting, Radiology 267 (1) (2013) 240–250.
[119] P.A. Marcovici, G.A. Taylor, Journal Club: structured radiology reports are more complete and more effective than unstructured reports, AJR Am. J. Roentgenol. 203 (6) (2014) 1265–1271.
[120] L.H. Schwartz, D.M. Panicek, A.R. Berk, Y. Li, H. Hricak, Improving communication of diagnostic radiology findings through structured reporting, Radiology 260 (1) (2011) 174–181.
[121] B. Bryl, S. Merrix, D. Proud, A. Marin, A. Byrne, J. Duckers, Are we missing the opportunity to measure muscle mass on computed tomography thorax? J. Thorac. Imag. 36 (2) (2021) W32–W33.
[122] M.A. Mall, N. Mayer-Hamblett, S.M. Rowe, Cystic fibrosis: emergence of highly effective targeted therapeutics and potential clinical implications, Am. J. Respir. Crit. Care Med. 201 (10) (2020) 1193–1208.
[123] T. Flohr, M. Petersilka, A. Henning, S. Ulzheimer, J. Ferda, B. Schmidt, Photon-counting CT review, Phys. Med. 79 (2020) 126–136.
[124] M.J. Willemink, M. Persson, A. Pourmorteza, N.J. Pelc, D. Fleischmann, Photon-counting CT: technical principles and clinical prospects, Radiology 289 (2) (2018) 293–312.
[125] M.M. Woeltjen, J.H. Niehoff, A.E. Michael, S. Horstmeier, C. Moenninghoff, J. Borggrefe, et al., Low-dose high-resolution photon-counting CT of the lung: radiation dose and image quality in the clinical routine, Diagnostics 12 (6) (2022).
[126] V. Van Ballaer, A. Dubbeldam, E. Muscogiuri, L. Cockmartin, H. Bosmans, W. Coudyzer, et al., Impact of ultra-high-resolution imaging of the lungs on perceived diagnostic image quality using photon-counting CT, Eur. Radiol. 34 (3) (2024) 1895–1904.
[127] M. Remy-Jardin, A. Hutt, T. Flohr, J.B. Faivre, P. Felloni, S. Khung, et al., Ultra-high-resolution photon-counting CT imaging of the chest: a new era for morphology and function, Investig. Radiol. 58 (7) (2023) 482–487.
[128] M.J. Siegel, S.M. Bugenhagen, A. Sanchez, S. Kim, A. Abadia, J.C. Ramirez-Giraldo, Comparison of radiation dose and image quality of pediatric high-resolution chest CT between photon-counting detector CT and energy-integrated detector CT: a matched study, AJR Am. J. Roentgenol. 221 (3) (2023) 363–371.
[129] S. Dettmer, T. Werncke, V.N. Mitkovska, T. Brod, O. Joean, J. Vogel-Claussen, et al., Photon counting computed tomography with the radiation dose of a chest X-ray: feasibility and diagnostic yield, Respiration 103 (2) (2024) 88–94.
[130] Bekanntmachung der aktualisierten diagnostischen Referenzwerte für diagnostische und interventionelle Röntgenuntersuchungen, Available from: https://www.bfs.de/SharedDocs/Downloads/BfS/DE/fachinfo/ion/drw-roentgen.pdf;jsessionid=070FF08F6A94FB48F42F3C6FADB51807.1_cid381?_blob=publicationFile&v=13, 2022.
[131] P.W. O'Regan, N.E. Stevens, N. Logan, D.J. Ryan, M.M. Maher, Paediatric thoracic imaging in cystic fibrosis in the era of cystic fibrosis transmembrane conductance regulator modulation, Children 11 (2) (2024).
[132] WHO, Communicating radiation risks in paediatric imaging, Available from: https://www.who.int/publications-detail-redirect/978924151034, 2016.