



Guidance for chest-CT in children and adults with cystic fibrosis: A European perspective (part 2: special situations/respiratory complications)

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Abbreviations: ABPA, Allergic Bronchopulmonary Aspergillosis; Af, *Aspergillus fumigatus*; BAE, Bronchial Artery Embolisation; BAL, bronchoalveolar lavage; CF, Cystic Fibrosis; CFF, Cystic Fibrosis Foundation; CR, Chest Radiography; CT, Computed Tomography; ECFS, European Cystic Fibrosis Society; ECFS-CTN ECFS, Clinical Trials Network; HAM, hyper-attenuated mucus; LUS, lung ultrasound; MAC, *Mycobacterium avium* Complex; MABSC, *Mycobacterium abscessus* Complex; MBW, Multiple Breath Washout; MDCTA, Multidetector CT Angiography; MSSA, methicillin-sensitive staphylococcus aureus; NTMD, non-tuberculous mycobacterial disease; Pa, *Pseudomonas aeruginosa*; PNT, pneumothorax; ppFEV₁, percent predicted; PSP, primary spontaneous pneumothorax; PwCF, People with Cystic Fibrosis.

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ABSTRACT

This manuscript covers guidance proposed by The European Cystic Fibrosis Society Clinical Trials Network (ECFS-CTN) for the use of chest computed tomography (CT) scans in special situations such as haemoptysis, aspergillus-related lung disease, non-tuberculous mycobacterial (NTM) infection and pneumothorax in people with CF. Guidance development followed a three-step process.

The most important questions were first identified via anonymous online survey, then a comprehensive literature search was made to support a first set of statements. They were finally validated by a Delphi process to obtain collective opinion of participants. Consensus supported the use of chest CT in the presence of haemoptysis, Allergic Bronchopulmonary Aspergillosis, and NTM-positive cultures (>2 separate expectorated sputum samples, or 1 positive bronchoscopic result). The ultimate responsibility for justifying a chest CT scan lies with the healthcare professionals directly involved in patient management.

1. Introduction

This document, produced by the Standardisation Committee of the European Cystic Fibrosis Society Clinical Trials Network (ECFS-CTN) provides guidance on monitoring strategies for chest computed tomography (CT) scans in special situations and respiratory complications in people with cystic fibrosis (PwCF), intended for clinicians, radiologists, researchers, and patients. It is intended to complement the ECFS-CTN guidance on monitoring strategies for chest CT [1].

These recommendations primarily address non-tuberculous mycobacterial disease (NTMD), allergic bronchopulmonary aspergillosis (ABPA), haemoptysis, pneumothorax and atelectasis.

2. Methodology

This practical European guidance is based on: (1) an online survey conducted by the European Cystic Fibrosis Society Clinical Trials Network (ECFS-CTN), (2) a review of the available literature, and (3) a Delphi validation process, as previously described [1].

Briefly, a multidisciplinary panel of 7 paediatric and 5 adult pulmonologists, 1 paediatric and 4 adult radiologists, 1 standardization specialist - all experienced in CF care - and a study methodologist were involved in coordinating the selection and definition of the clinical and research questions.

Our Delphi process encompassed several iterative rounds, three rounds in total were completed on the following dates: Delphi Round 1: 27th November 2023 (n = 19); Delphi Round 2: 23rd February 2024 (n = 18); Delphi Round 3: 3rd March 2024 (n = 24).

Delphi recommendations were based on a careful review of the literature and on surveys and consensus discussions with experts in the field. An initial survey was sent to all 57 ECFS-CTN CF centers. A total of 43 responses were received for the paediatric sub-group and 22 for the adult sub-group. The Delphi committee drew up a list of recommendations based on either evidence or expert opinion on the clinical question. Online Delphi rounds were then used to identify agreement and disagreement, which were voted on using a nine-point Likert scale: strongly disagree (score 1), disagree (score 3), neutral (score 5), agree (score 7) and strongly agree (score 9) [2]. As per recommendations we excluded a maximum of one extreme value per 15 members if no value was missing e.g. the minimal value if the median was >5 or the maximal value if the median was ≤5. Rounds were ended when consensus was reached or when it was clear that no agreement could be reached: stable disagreement >2 rounds or when a maximum of 3 rounds were completed [3]. Round 1, round 2 and round 3 comprised 19, 14 and 24 participants, respectively. Subsequent expert review was completed 28th March 2024. A medical statistician with experience in the Delphi

process was part of the methodology team and part of the authorship team.

In the ECFS-CTN online survey, participants considered the following conditions to be the most important regarding the use of CT scans for the diagnosis of respiratory complications in CF (in descending order of frequency): In CHILDREN (see Fig. 1) : Haemoptysis, Persistent alveolar consolidation, Allergic Broncho-Pulmonary Aspergillosis (ABPA), Bronchiectasis and Pneumothorax; and in ADULTS (see Fig. 2) : Non-tuberculous mycobacteria (NTM) infection, ABPA/Aspergillus infection, Haemoptysis, Pneumothorax, Persistent atelectasis.

The databases used for the literature search were 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-analyses, 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 (USPSTF) system for assessing the quality of evidence and strength of recommendations.

2.1. Non-tuberculous mycobacteria (NTM)

Non-tuberculous mycobacteria (NTM) are opportunistic pathogens of growing concern for people with cystic fibrosis (PwCF), as they can significantly reduce quality of life and lead to poor outcomes [4]. They are commonly found in water and soil. While previously thought to be purely environmental, emerging evidence suggests possible person-to-person transmission in CF centers, although this remains debated [5,6]. The most common species affecting PwCF are *Mycobacterium avium* complex (MAC) and *Mycobacterium abscessus* complex (MABSC).

CF is a risk factor for NTM infection, with prevalence varying geographically and by screening practices, typically 1–7% in the UK and Europe [7–10] and up to 14% in the USA [11]. Rates appear to be increasing due to improved detection, longer life expectancy, and treatment factors such as antibiotic use [12]. The impact of CFTR modulators is unclear: some studies show reduced incidence, while others report persistence of NTM [13–16].

Diagnosing NTM pulmonary disease (NTM-PD) is complex. It requires a combination of clinical symptoms, radiological findings (e.g. nodules, cavitation, bronchiectasis), and microbiological evidence (e.g. multiple positive sputum cultures). A positive culture alone does not confirm disease. Diagnosis follows established guidelines (e.g. ATS/ERS and CF Foundation/ECFS) [17–19].

The 2007 ATS/ERS/ESCMID/IDSA Guidelines were the first to recommend the introduction of annual screening for NTM in PwCF. It

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recommends that all expectorating patients should be screened for NTM regularly, at least once a year, and that all PwCF, including children, should be screened for NTM during periods of clinical deterioration that does not respond to treatment for non-NTM pathogens [17] (A,II). The CFF (Cystic Fibrosis Foundation) recommends that cultures for NTM should be performed annually in spontaneously expectorating individuals with a stable clinical course [18]. As stated in the CFF/ECFS Guidelines, the diagnostic criteria for NTM-PD in PwCF should be the same as in patients without CF, although they may be more difficult to apply due to the underlying disease and concomitant infection with overlapping clinical features and radiological manifestations [17,19] (C, III). Therefore, chest radiography (CR) in the context of CF-related lung disease is unlikely to be useful for the investigation of NTM-PD. Radiological changes that support a diagnosis of NTM-PD include: cavitation, inflammatory nodules, and new ‘tree-in-bud’ opacities (especially in areas of mild underlying bronchiectasis), although the latter two features are less specific as they are associated with usual bacterial infection in CF [20]. These manifestations can also be attributed to common bacterial infection and interpretation of radiological findings can be difficult in CF [6].

For this section, we conducted a literature search in the PUBMED database using the MeSH terms “non-tuberculous mycobacteria”, “NTM”, “chest tomography (CT) scan”, “radiology” and “cystic fibrosis” and found a total of 23 studies. Despite our literature search, some questions regarding the use of CT scans in the treatment of NTM-PD remain unanswered due to variability in practice and equipment between CF centers. Questions were included in the online survey to assess practice in relation to NTM management.

2.1.1. Potential role of CT scans in the management of NTM airway infections in PwCF

The CFF recommends at least annual NTM sputum culture screenings for patients with CF [19]. It is important to emphasise that the recommended screening for NTM infection refers to the collection of samples from individuals in whom there is no clinical or radiological suspicion of NTM infection and should be distinguished from strategies to investigate and diagnose NTM disease in symptomatic patients. Therefore, once NTM species and subspecies have been isolated and identified, their clinical relevance should be carefully considered.

Indeed, one area of interest is the assessment of the clinical relevance of an NTM-positive sputum culture and the early detection of the first signs of lung disease. Recently, a single-centre cross-sectional study investigated the characteristics of chest CT in patients with NTM-PD

compared with asymptomatic NTM infection [21]. In the first group, the airways were more severely affected in terms of the severity of bronchiectasis. In addition, the presence of cavitation was only detected in the NTM-PD group, indicating that chest CT features may be useful as a diagnostic and potentially prognostic biomarker for the risk of transition from NTM colonisation to PD [21].

In contrast to these findings, in a retrospective study, Lam et al. evaluated 13 PwCF with a positive sputum culture for mycobacterium abscessus, 9 of whom met the ATS/IDSA criteria for NTM-PD and appropriate treatment was initiated [22]. Bronchiectasis, mucus plugging, and consolidation were common findings. However, these findings are also common in CF lung disease, and no specific radiological signature has been identified to reliably distinguish NTM lung disease. For this reason, the lack of specific CT findings at the time of initial infection should not reassure CF clinicians about the possibility of disease progression.

In our online survey, we investigated the use of chest CT in patients with NTM-positive sputum cultures who were not diagnosed with NTM-PD. In adult CF patients, 67% of physicians would consider a chest CT scan in all patients to monitor for possible progression; 24% would consider a CT scan only in clinically worsening patients; 9% would not consider a chest CT scan in this situation. In contrast, in a paediatric CF population, 42% of physicians would consider a CT scan of the chest in all patients, 40% would only consider a CT scan if the condition worsened and 18% would not perform a CT scan. We also enquired about the frequency of radiological follow-up in CF patients with an NTM-positive sputum culture, and consensus was not reached.

Delphi Statement n°	Type of statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
1	CT scanning is the gold standard for radiological diagnosis and follow-up of NTM-PD, even though there are overlaps between the radiological findings of NTM-PD and CF.	GRADE C*	2+	9 [6–9] APPROPRIATE (Relatively Strong Agreement)

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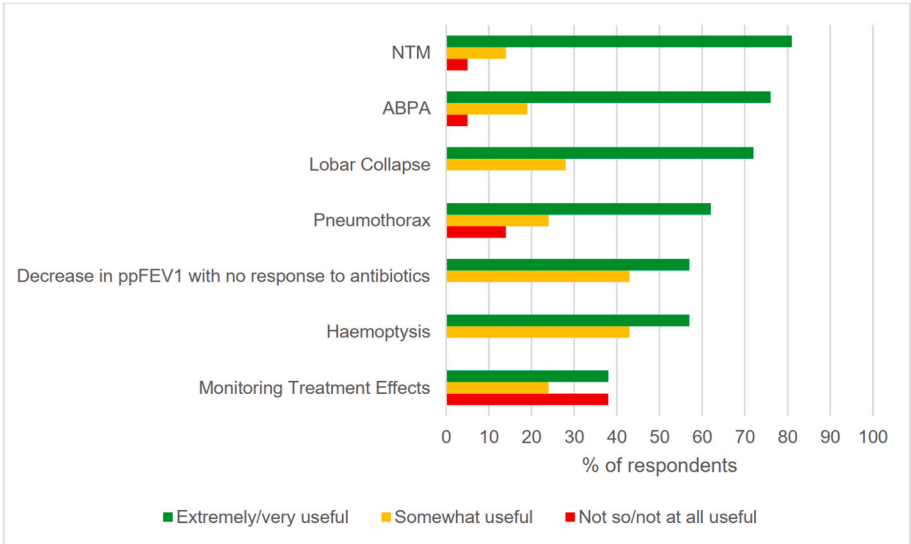


Fig. 1. Perceived usefulness of chest CT-scans in children, according to the clinical scenario (ECFS-CTN survey).

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Delphi Statement n°	Type of statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
2	In asymptomatic patients of any age with a first NTM-positive culture, a (baseline) CT scan can be considered.	GRADE D**	4	9 [2-9] UNCERTAIN (No Consensus)
3	In asymptomatic patients of any age with multiple NTM-positive cultures, a (baseline) CT scan may be considered.	GRADE D**	4	9 [7-9] APPROPRIATE (Strong Agreement)

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

2.1.2. Role of chest CT-scans in NTM-PD in PwCF

As previously reported, diagnostic criteria for NTM lung disease include clinical, microbiological, and radiological findings [17,19]. Although not specifically defined for pwCF, these criteria are generally applied to this category of patients. Diagnosing NTM disease in PwCF is challenging because CF-related symptoms and imaging abnormalities overlap with those of NTM, which lowers the specificity of radiological features typically associated with NTM-PD [23]. Even when obvious nodular or cavitary lesions can be recognised on a chest radiography, chest CT remains the standard for diagnosis [19].

NTM pulmonary disease demonstrates two principal radiological phenotypes: fibrocavitary and nodular-bronchiectatic disease. Fibrocavitary disease is most commonly associated with *Mycobacterium avium* complex (MAC), whereas *Mycobacterium abscessus* is more frequently linked to nodular-bronchiectatic patterns, particularly in people with cystic fibrosis. Although *M. abscessus* is associated with more severe clinical outcomes and accelerated lung function decline, it is less typically associated with classical fibrocavitary disease [24,25].

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Author/year	CT appearance of NTM
Author/year	CT appearance of NTM
Kim 2005	On initial CT scans, bronchiectasis and nodules were the most frequent findings of NTM infection in all three groups (classified by CF mutation: n=15 single CF mutation; n=13 abnormal AAT; n-3 heterozygous CF mutation and heterozygous AAT phenotype). Prevalence of nodules was slightly lower in the CF group, and the prevalence of linear scarring was greater in the AAT group.
Garcia 2022	NTM-PD patients had: - greater median number of lung lobes affected by bronchiectatic airways, - greater frequency of bronchiectatic airways - greater frequency of cystic bronchiectasis vs NTM-C - Cavitation of lung parenchyma was a radiographic feature solely associated with NTM-PD: mean cavity size was 3.13 ± 1.58 cm with a mean wall thickness of 0.58 ± 0.42 cm. Nodules were common, occurring in 77/84 (91.7%) of all patients. Use of the modified Reiff score to quantify the severity of bronchiectasis showed significantly greater (worse) scores for NTM-PD than NTM-C patients.
Lam 2028	All n=13 <i>Mycobacteria abscessus</i> CF patients had: - bronchiectasis and mucus plugging, with 10 of 13 (76.9%) in an upper lobe distribution - Consolidation was seen in 12 of 13 (92.3%) patients - 8 (61.5%) patients had nodules - 5 (38.5%) with cavitation - 3 (23.1%) had lung cysts. In patients with consolidation: - 9 out of 13 (69.2%) having upper lobe predominant consolidation - three patients (23.7%) with ground glass opacities No sig difference between patients with colonisation versus disease.
Kwak 2016	Correct NTM-PD diagnoses: - Tree-in-bud pattern - consolidation and - atelectasis Presence of pleural effusion led to false diagnoses.
Koh 2017	Fibrocavitary lesions are predominantly upper lobe in MAC (<i>Mycobacterium avium</i> complex)
Hendrix 2022	<i>Mycobacterium abscessus</i> is more frequently linked with nodular/bronchiectatic patterns rather than classic fibrocavitary disease

*NTM-pulmonary disease meeting ATS/IDSA guidelines for treatment; **NTM-colonised but not meeting ATS/IDSA criteria for NTM treatment.

NTM-PD typically manifests as either a nodular-bronchiectatic or a fibrocavitary pattern. Chest CT images (panels A1 and A2) show the

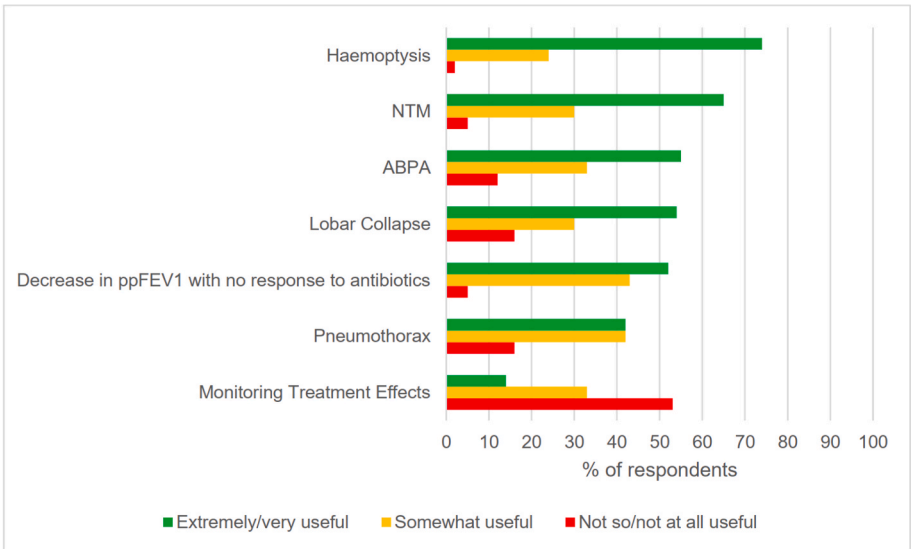


Fig. 2. Perceived usefulness of chest CT-scans in adults, according to the clinical scenario. (ECFS-CTN survey).

radiological manifestations in a 35-year-old female PwCF (F508del/D110H), chronically infected with methicillin-sensitive *Staphylococcus aureus* (MSSA) and normal lung function (ppFEV₁ 100) of a nodular-bronchiectatic pulmonary disease by *Mycobacterium abscessus*. Focal segmental or subsegmental varicose or cylindrical bronchiectasis (red arrows) might be visible, sometimes associated with small nodules, bronchial wall thickening, mucus plugs and/or centrilobular “tree-in-bud” pattern (green circles).

The fibrocavitary pattern is characterised by larger cavitary lesions, often surrounded by adjacent consolidations and area of parenchymal fibrotic distortion with volume loss, as shown in panel B1, B2, B3 (50-year-old female PwCF, F508del/D1152H, ppFEV₁ 65, no chronic infection, with *Myc. Abscessus bolletii* pulmonary disease) and C1 (58-year-old female PwCF, F508del/D1152H, ppFEV₁ 30, PA and *Achromobacter* chronic infections, with MAC pulmonary disease).

In PwCF radiological manifestations of NTM-PD may be difficult to distinguish from CF pulmonary disease, as shown in pictures D1 and D2 (46-year-old male PwCF, F508del/711+3A > G, ppFEV₁ 40, PA and MSSA chronic infection, with *M. a. abscessus* pulmonary disease) (see Fig. 3).

None of the patients whose CT scans are shown in Fig. 3 were treated with CFTR modulator at the time the images were acquired.

Given the poor outcomes for *M. abscessus* despite intensive, long-term, multidisciplinary and burdensome therapy with frequent treatment-related side effects [26], treatment decisions should be individualized based on host factors, disease severity and likelihood of disease progression. Initiation of antimicrobial therapy is recommended for patients with a positive smear with acid-fast bacilli or cavitary lesions [19]. Regarding the nodular/bronchiectatic form of NTM disease, the

available literature is inconclusive [27].

NTM species responsible should also be considered when deciding whether to initiate a specific treatment. Identifying the specific NTM species is crucial because treatment regimens, drug susceptibility, and clinical prognosis can vary significantly between species [28].

Even in cases where treatment is initiated, there is a lack of data on how patients with confirmed NTM-PD should be followed up with imaging to assess response to treatment. According to the CFF/ECFS Guidelines, chest CT should be performed in PwCF with confirmed NTM-PD shortly before treatment initiation and at the end of treatment (recommendation #37. 94% consensus) [19]. A follow-up HRCT of the chest is also recommended at end of treatment, e.g., 1 year after treatment initiation. However, there is very little data to support these recommendations.

Moreover, different NTM species may have different outcomes and therefore CT scan interpretation may require a combination approach of the radiographic patterns [28,29].

In our survey in adult CF patients with NTM-PD, 76% of physicians would use a chest CT scan to monitor response to treatment, with 65% performing this scan around 6-12 months after starting treatment. In paediatric CF patients, 60% of physicians would use a chest CT scan to monitor response to treatment, although there is no consensus on the timing.

Delphi Statement n°	Type of statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
4	A follow-up CT scan can be	GRADE D**	4	9 [7-9] APPROPRIATE (Relatively)

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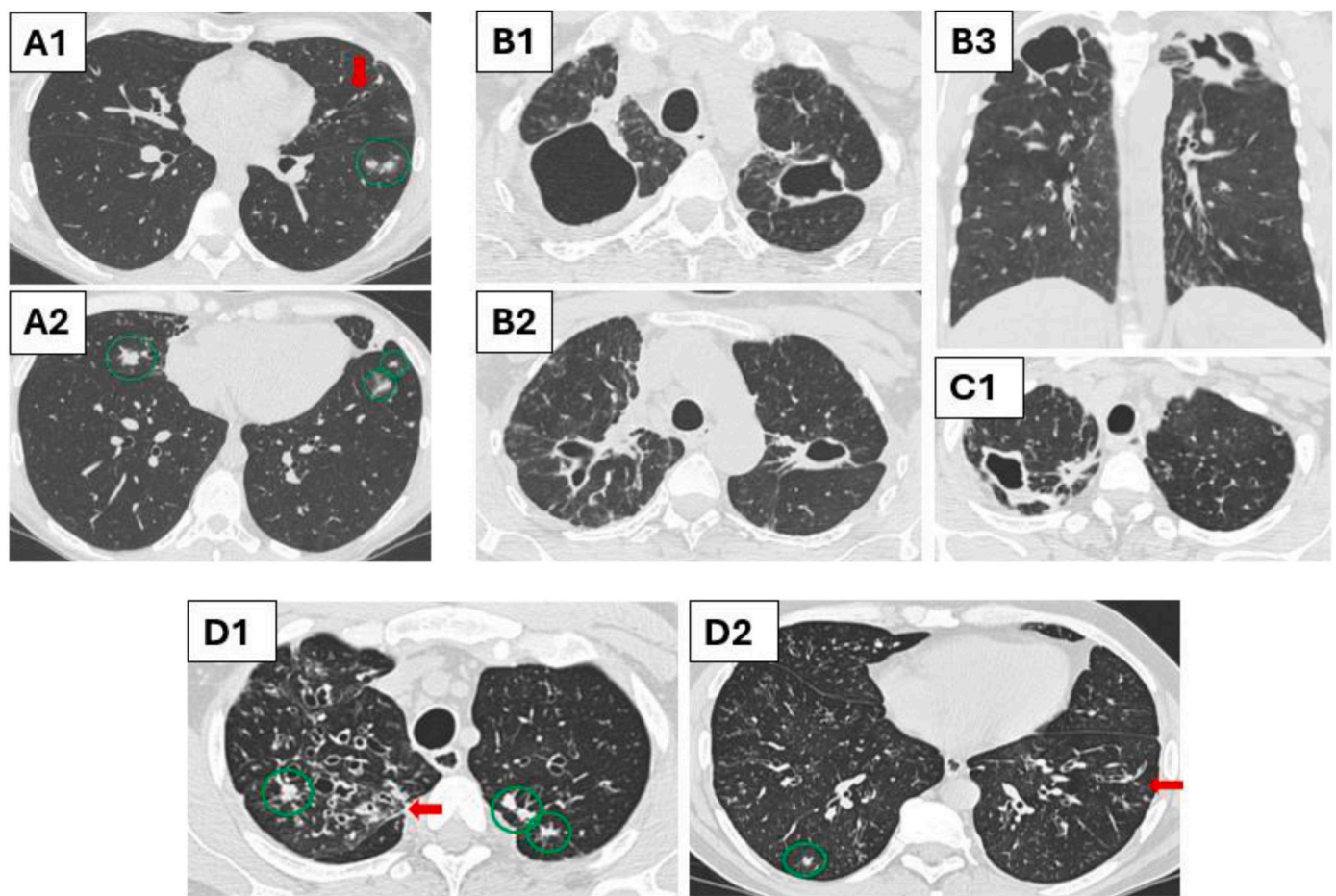


Fig. 3. Chest CT-scans in non-tuberculous mycobacteria (Courtesy Luca Riberi & Barbara Messore).

(continued)

Delphi Statement n°	Type of statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
	considered 6–12 months after starting treatment for NTM-PD in PwCF.			Strong Agreement)

**RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

2.2. Aspergillus-related diseases

The most clinically relevant species associated with human lung disease is *Aspergillus fumigatus* (Af). In CF, the CFTR protein defect leads to abnormal sputum viscosity and impaired mucociliary clearance, which may favor fungal colonisation [30]. Several reports suggest that Af, in addition to bacterial infections, may contribute significantly to worsening symptoms, deterioration of lung function and exacerbations and progression of CF lung disease in children. Nevertheless, it is difficult to distinguish Af-related disease from other clinical conditions, and imaging of the lungs using chest CT can help to differentiate Af-related from other microorganism-induced damage to aid clinical management and treatment [31]. However, diagnosis always relies on a combination of symptoms, microbiological and biological samples, immunology and imaging. The specificity of imaging techniques may vary depending on the underlying inflammation.

Firstly, colonisation by Af corresponds to the presence of fungi in laboratory cultures of the respiratory tract without clinical symptoms or a deterioration in lung function. No specific structural changes have been described in the literature to date. However, it has been shown that Af colonisation is associated with co-infection with *Pseudomonas aeruginosa* [32]. In addition, patients colonised with Af may show more severe bronchiectasis and air trapping, although causality could not be established [33]. HRCT scores, when combined with serological markers (IgG, IgM, galactomannan), may improve the diagnostic accuracy for active Af infection and help guide further invasive evaluation [34].

Secondly, Af bronchitis or bronchiolitis corresponds to an active bronchial or bronchiolar active infectious inflammation of the airways. Direct invasion of the bronchial wall layer by Af has been demonstrated in pathological specimens [31,35]. Thus, on CT scans, wall thickening and centrilobular nodules are the most frequently encountered manifestations, with surrounding consolidations, as the disease can spread from the airways to the adjacent lung parenchyma [36].

Thirdly, a fungal ball (aspergilloma) can develop in a pre-existing lung cavity. In CF, these cavities may correspond to an abscess, a cyst, or sometimes severely dilated bronchi. Two CT signs can help to recognise the presence of a fungal ball. The Monod sign indicates the presence of air in the vicinity of the fungal ball. In addition, the fungus is likely to be mobile in the cavity, and an additional CT scan in the prone position can increase diagnostic certainty by showing gravity-dependent displacement of the fungal ball [37].

Fourth, allergic bronchopulmonary aspergillosis (ABPA) corresponds to allergic inflammation due to exposure to Af. Recent publications agree that chest CT, not CR, plays a key role for diagnosing ABPA in PwCF [38] and outside of CF [39]. Most CT signs are non-specific, such as the presence of bronchiectasis in the central airways, mucus plugs, centrilobular nodules and consolidations. In the past literature, the term “infiltrate” used to describe migrating pulmonary ABPA-related lung opacities [40]; however, recently published glossary of terms by the Fleischner Society considers that the term “infiltrate” is obsolete and should be replaced by “fleeting opacities” [41]. Distinguishing ABPA from other pulmonary infectious processes is important because ABPA requires specific therapeutic management with corticosteroids and sometimes antifungals or biologics, but not antibiotics. However, as

most of the CT signs mentioned above are non-specific, repeated imaging during treatment is recommended [42]. It is worth mentioning that hyper-attenuated mucus (HAM) has been shown to be a highly specific CT sign of ABPA. In ABPA patients with central mucus, HAM can be recognised as mucus with a higher attenuation than the paraspinal musculature and/or a measured attenuation between 70 and 150 Hounsfield units (see Fig. 4). Recently, The International Society for Human and Animal Mycology (ISHAM) Group has proposed that the presence of this CT sign can confirm the disease, even if patients would not fulfill all the other immunological or biological criteria [39]. However, in CF, several cautions may be considered. Indeed, the ISHAM group recommend using CT standard doses of 3-7 mSv, while ultralow CT doses (approximately 0.08 mSv) have been recommended [1]. Thus, the reduction of CT doses may modify the diagnostic attenuation values. For instance, by using a ratio higher than the paraspinal muscle with low dose CTs, a 100% specificity was reached for a ratio >1.8 instead of >1 in CF [43].

Nevertheless, the sensitivity of HAM in CF may be higher than in non-CF conditions and was found to be around 70% [44]. Also, in expert centers using MRI in the routine management of CF, especially when repetition of imaging tests is necessary [45,46], an alternative to HAM at CT is the so-called IMIS (Inverted Mucoid Impaction Signal) sign [47]. Of note, IMIS is based on a characterization of mucus impaction itself without the need for an extrapulmonary muscular reference and has demonstrated a similar specificity to CT [43].

Finally, annual assessment of ABPA status is one of the international recommendations for the annual routine examination of CF patients, while imaging is part of the diagnostic criteria [40,42].

In our survey, the performance of a CT scan of the chest in the case of Aspergillus-related disease in PwCF was considered useful by 95% of adult physicians and in 89% of paediatricians.

In summary, there is recent evidence that CT is more sensitive and specific than CR for detecting Aspergillus-related lung disease, including in CF [40,48,49]. Agarwal (2024), using a Delphi Expert Consensus approach, recommended a thin-section chest CT at baseline to identify and characterise bronchiectasis, mucus plugging, HAM and other abnormalities in patients with suspected ABPA. Whilst Laurent (2024) and Schwarz (2024) suggest that CT is more sensitive and specific than CR for detecting Aspergillus-related lung disease, including in CF [40,48,49]. CT appears integral for diagnosis, follow up, and treatment evaluation [50,51]. Also, CT is essential for identifying structural changes associated with Af infection or ABPA in CF, and it is used to monitor disease progression and response to therapy [49,52]. New CT technologies with CT dose reduction and AI-based image analysis are in process which may further improve sensitivity for detecting fungal-related changes [52,53].

2.3. Haemoptysis

Haemoptysis is a worrying complication that can occur in PwCF [54]. Although the clinical course of the disease is changing in the era of CFTR modulator therapy, haemoptysis occurs in a significant percentage of PwCF. Studies performed prior to CFTR modulator triple combinations have estimated that approximately 9% of PwCF will experience haemoptysis within 5 years and 4-6% will experience at least one episode of massive haemoptysis during their lifetime [55,56], with an annual incidence of 0.9-1% [57] (see Fig. 5). In addition, around 25% of these patients experience a recurrence of haemoptysis [55,58]. However, more recent data show that the scenario is changing. In the 2023 CFF Patient Registry, the incidence of haemoptysis was 0.3% in PwCF <18 years of age and 2.7% in those ≥18 years of age, with an overall incidence of 1.7%. Specifically, massive haemoptysis occurred in less than 0.1% of PwCF <18 years, and in 0.3% ≥ 18 years, with an overall incidence of 0.2% [59]. The ECFS Patient Registry (ECFSR 2023) investigated the incidence of massive haemoptysis in patients who have never had a transplant. In those <18 years of age 44 cases (0.2 %)

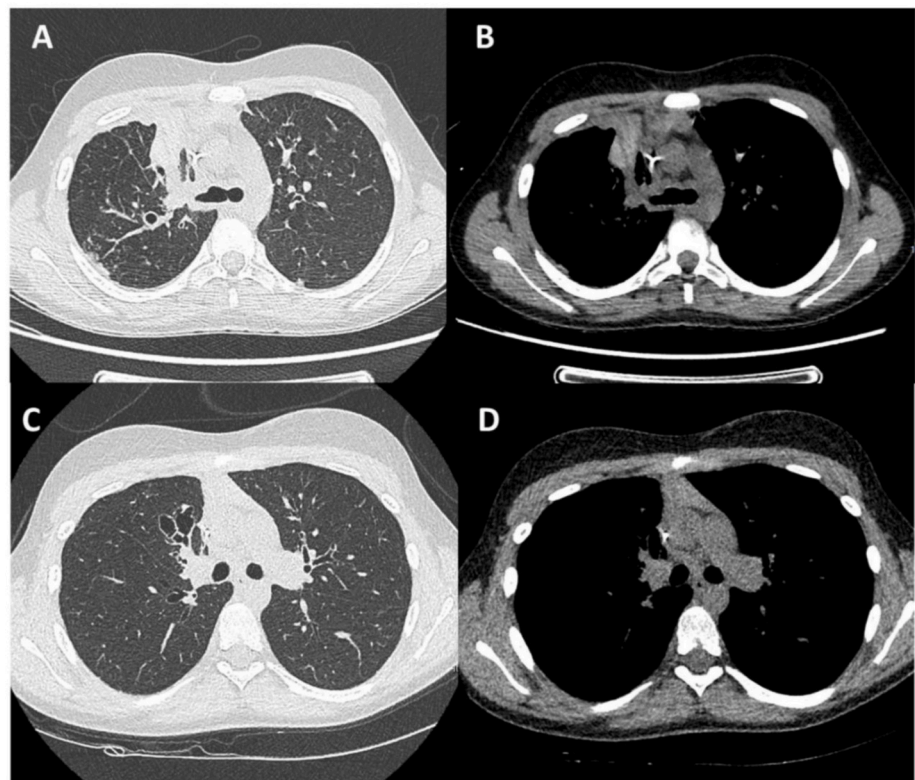


Fig. 4. CT of an adult case of Allergic Broncho-Pulmonary Aspergillosis (ABPA) (Courtesy Gael Dournes). During an acute exacerbation (A) with Hyper-attenuated mucus (HAM) in the right upper lobe (B). Residual bronchiectasis after treatment (C) with absence of HAM (D).

occurred, while in those ≥ 18 years of age 261 cases (1.0%) occurred [9].

Most of these events are classified as “scant” or “moderate” [60], although some “massive” episodes may occur. A study of a retrospective cohort of 1008 PwCF not treated with CFTR modulators estimated that 76% of events were mild and only 9% were massive [61]. Although rare, massive haemoptysis can be a potentially fatal complication of CF lung disease. It can also lead to a more rapid decline in lung function, an accelerated need for lung transplantation and increased mortality [62]. Older age and more severe lung disease have been shown to be risk factors [62].

The most recent published recommendations on haemoptysis are the 2010 CFF Guidelines [60]. The authors also questioned any potential benefit of a contrast-enhanced chest CT scan in this case. Despite a specific point that all patients with haemoptysis should have a CT angiography of the chest before bronchial artery embolisation (BAE), no agreement was reached. Some felt it was useful but not essential, while others felt it was a waste of valuable time. A subsequent paper by Hurt and Simmonds [63] suggested that new insights could be gained with the spread of multidetector CT angiography (MDCTA). This technique could allow a more detailed reconstruction of the anatomy of the bronchial arteries [64].

Today, clinical management and treatment protocols vary from centre to centre. In clinical practice MDCTA is frequently used to localise the bleeding site prior to BAE. It is also useful to provide a road map and anatomical details of the bronchial artery. The efficacy of MDCTA in detecting the bleeding vessel was estimated to be approximately 97.5% of cases in an adult non-CF cohort [65], with 90% of cases involving the bronchial arteries in the haemorrhage [57].

Sheppard et al. have discussed the management approaches to haemoptysis in young PwCF < 18 years, referencing the use of chest CT as part of the diagnostic and management algorithm, particularly for evaluating the extent of lung disease and guiding interventions such as BAE. Their institutional experience and literature review highlight the role of CT in classifying haemoptysis severity and informing

management decisions, in line with the US CFF Guidelines [66]. They categorised 53.3% of haemoptysis events as minor: 38.3% as moderate and 8.3% as major. In this cohort, MDCTA was used in 28.3% of cases, most of which (10 of 17) were for moderate haemoptysis [66]. The most common parenchymal abnormality was bronchiectasis followed by consolidation and the most common vascular finding was hypertrophy or tortuosity of the bronchial arteries. They suggested using MDCTA for moderate or massive haemoptysis in clinically stable patients or in patients with mild haemoptysis who do not respond to an initial medical approach [66].

In our survey, the performance of a CT scan of the chest in the case of haemoptysis in PwCF was considered useful by 100% of adult physicians and in 98% of paediatricians. In addition, haemoptysis was considered to be one of the most common respiratory complications for which a chest CT scan is performed.

Delphi Statement n°	Type of statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
5	A contrast-enhanced CT scan should guide indication for embolisation of the bronchial arteries in PwCF with clinically significant haemoptysis.	GRADE D**	4	9 [5-9] APPROPRIATE (Relatively Strong Agreement)

**RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

2.4. Pneumothorax

Pneumothorax (PNT) can cause significant short-term morbidity and

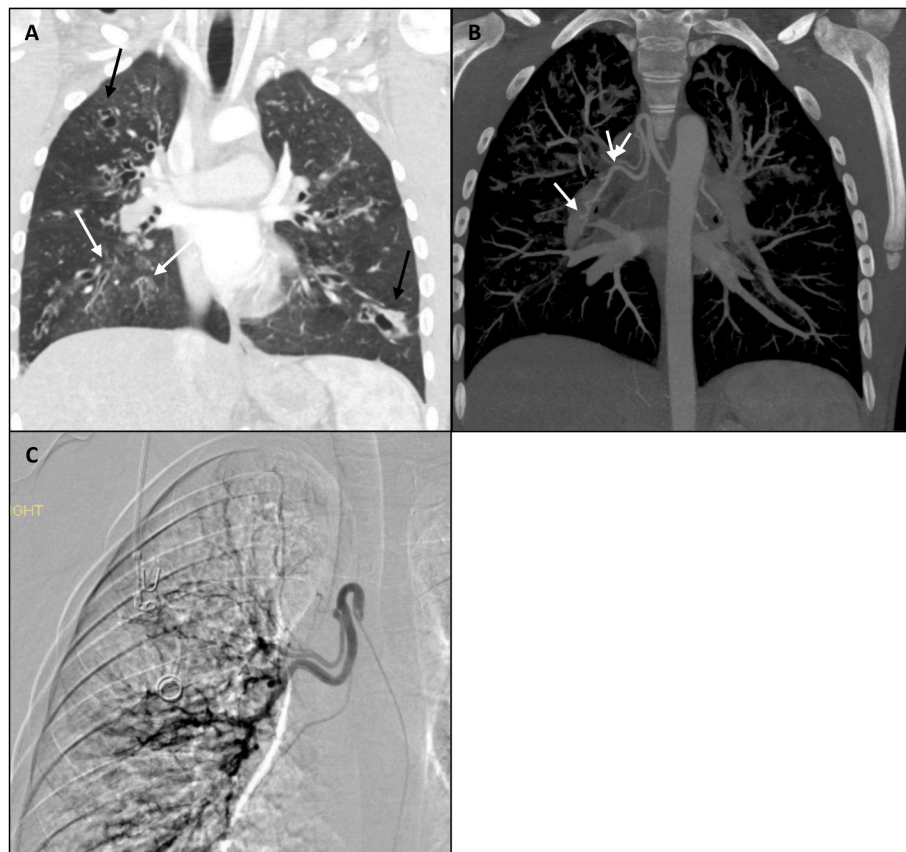


Fig. 5. CT of a 24-year-old PwCF presents with haemoptysis (Courtesy Michael Maher).

A. Coronal reconstructed image from CT thorax with IV contrast shows widespread bronchiectasis, seen in the right upper and left lower lobes (black arrows) on the current image. There are centrilobular ground glass nodules in the right lower lobe (white arrows), which were a new finding compared to the prior CT scan: these were considered likely to represent aspirated blood within the alveoli.

B. Coronal reconstructed maximal intensity projection (MIP) image of a CT angiogram shows significantly hypertrophied bronchial arteries on the right and left sides of the mediastinum. The largest hypertrophied bronchial artery coursed inferiorly through the right side of the mediastinum (arrows) towards the site of clustered centrilobular nodules in the right lower lobe. As the patient had persistent haemoptysis, the patient proceeded to selective bronchial angiography.

C. Selective bronchial angiography shows the anatomy of the hypertrophied bronchial artery on the right side of the mediastinum, which closely resembled the findings on CT angiography, performed 2 h previously. This was followed by catheter embolisation of the hypertrophied right bronchial artery, using 700–900 μm embospheres, with successful resolution of haemoptysis.

has also been associated with a poorer prognosis in subsequent years, presumably as it represents a marker of increased structural lung disease in PwCF. It occurs with an average annual incidence of 0.64%, and approximately 3.4% of PwCF will experience PNT during their lifetime [60] (see Fig. 6). PNT rarely occurs in children and adolescents, with prevalence increasing in older PwCF (median age 21 years) and those with advanced lung disease [67]. Recent data from the ECFS patient registry shows an incidence of PNT of 0.1% in PwCF <18 years and 0.2% in ≥ 18 years [9], with similar incidence in CFF registry data of less than 0.1% in PwCF <18-year-olds and an incidence of 0.1% in >18 -year-olds. Several risk factors have been associated with an increased risk of PNT: *P. aeruginosa*, *Burkholderia cepacia* and/or *Aspergillus* in sputum cultures, ppFEV₁ <30%, enteral tube feeding, pancreatic insufficiency, allergic bronchopulmonary aspergillosis, and massive haemoptysis. There was no increased incidence depending on gender. The clinical presentation ranges from mild or asymptomatic to a dramatic course.

The diagnosis of PNT is straightforward in the presence of suggestive symptoms or signs and can be easily made in most cases by a conventional CR. Other tests include a lung ultrasound (LUS) or a chest CT scan. LUS has recently become well established in the diagnosis of PNT in emergencies in non-CF trauma and critically ill patients. In a meta-analysis, the pooled sensitivity and specificity were 0.88 and 0.99 for ultrasonography and 0.52 and 1.00 for CR [68]. These studies showed a significantly higher diagnostic yield with LUS than with CR, using CT as

the reference examination [69]. Although LUS is extremely useful in the diagnosis of PNT, it should not be the only examination used to assess its extent and treatment [70].

In the 2023 guidelines on primary spontaneous pneumothorax (PSP) in non-CF patients, it is recommended that a low-dose CT scan of the chest should be performed after the first-line X-ray examination if diagnostic doubts persist [70]. Although chest CT is superior to CR in diagnosing secondary spontaneous pneumothorax, assessing its size and ruling out other differential diagnoses [71], the authors still consider it a second-line tool for assessment.

In PwCF while chest CT remains the most sensitive modality for detecting structural lung disease in CF, its routine use for pneumothorax is not universally recommended due to concerns about cumulative radiation exposure, especially in younger patients [52,72,73].

Thus, for the initial diagnosis and management of pneumothorax in CF, CR remains the first-line imaging modality [74]. CT is generally reserved for cases where the diagnosis is uncertain, when complications are suspected, or in the setting of recurrent or persistent PNT where structural abnormalities such as blebs or bullae may influence management decisions e.g. surgical interventions. Indeed, the diagnosis of PNT can be challenging in patients with advanced CF lung disease. Pleural adhesions are common, and lung collapse is therefore often uneven. The presence of pulmonary cysts and bullae [75], which can often be large and extensive, further complicates the assessment of the

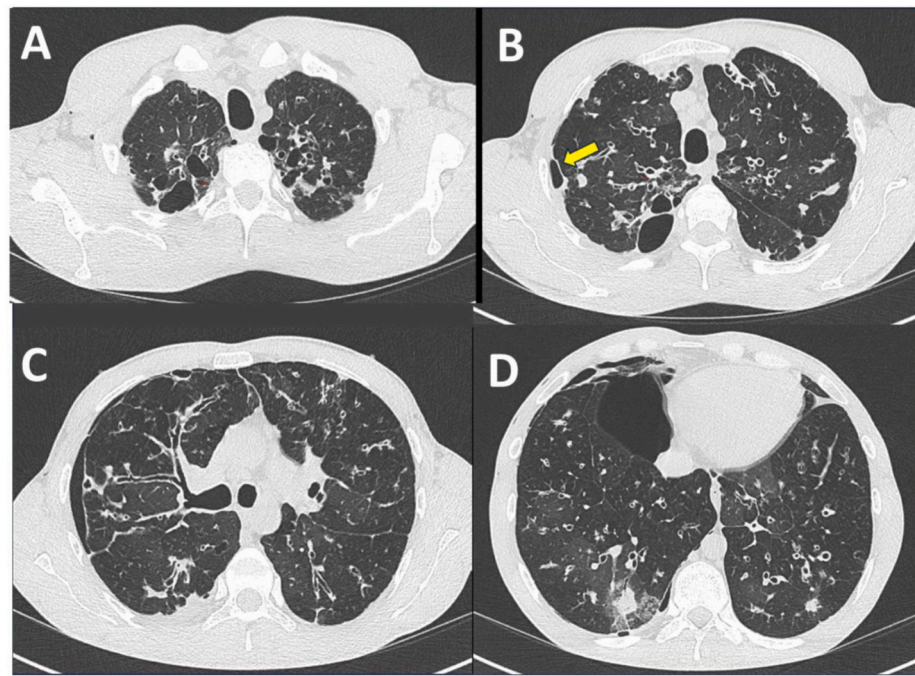


Fig. 6. Chest-CT-Scan in a 27-year-old male patient (Courtesy Dr Julie Macey, Bordeaux, France)
Panel A shows cystic lung disease in both upper lobes. In panel B, the cystic lesion indicated by the arrow is in fact the upper part of a pneumothorax, as seen in the lower panel C. A right paracardiac extension of the pneumothorax is seen in panel D.

extent of PNT or, in some cases, falsely suggests PNT. In patients with severe bullous lung disease, CT scanning may spare the patient an unnecessary and potentially dangerous chest tube insertion [76]. The diagnosis of PNT in PwCF can also be difficult in unusual cases (i.e. atypical localisation and small size) or when the CR is obscured by surgical emphysema [76]. The CT scan is the ultimate investigation to exclude or confirm the diagnosis of PNT. Therefore, a low suspicion threshold for performing a CT scan of the chest is justified in cases of suspected PNT. A CT scan allows a more accurate assessment of the size of a PNT to select appropriate treatment, determine a site for chest tube insertion and confirm resolution after treatment. However, CT scan should not delay the treatment of patients with acute respiratory failure, as these patients may require urgent CT scan for chest tube insertion [67, 77,78].

The treatment of PNT in PwCF is based on retrospective data and expert opinion, as there are no clinical trials to guide treatment. The 4 main factors that influence treatment of a PNT outside of CF are: 1) size of PNT, 2) severity of disease, 3) clinical stability of the patient, and 4) first or recurrent PNT [77]. It is generally recognised that ‘small’ is a PNT of 2 cm and “large” is a PNT of >2 cm in adults, measured along the axillary line between the lung margin and the chest wall at the level of the hilum [76]. Some other authors recommend the use of a cut-off of 3 cm from the dome [74]. The American recommendations state that: ‘A chest tube should be placed in patients with a large pneumothorax, and a chest tube should be placed in patients with a small PNT if there is clinical instability’ [60]. The British Thoracic Society’s 2023 Guidelines for pleural disease recommend a symptom-orientated approach, ‘Conservative treatment may be considered for the management of minimally symptomatic (i.e. no significant pain or breathlessness and no physiological impairment) or asymptomatic primary spontaneous PNT in adults regardless of size’ [79]. However, if a patient has high-risk presentation, such as tension PNT, significant hypoxia, bilateral PNT or underlying lung disease, intercostal drainage or needle aspiration may need to be considered if the procedure is safe. Medical treatment should usually include non-specific measures (chest physiotherapy and airway clearance, antibiotics, nutrition, and pain management). Surgical pleurodesis may be

considered for recurrent pneumothorax using techniques that do not lead to future contraindications for lung transplantation.

CFTR modulator therapies may provide additional benefit in terms of reducing pneumothoraces requiring chest drainage. As PNT is most common in PwCF with advanced lung disease, reducing the development of structural lung abnormalities using CFTR modulator therapy earlier in life may reduce the incidence of future PNTs.

In summary, chest CT is not routinely indicated for all CF patients with pneumothorax. Its use should be individualized and restricted to selected cases, primarily for complex or recurrent cases, or in post-surgical (post-video-assisted thoracic surgery) refractory or recurrent PNT where additional anatomical detail will impact management.

Delphi Statement n°	Type of statement	Strength of recommendation	Quality of evidence	Median [Range] Delphi Score
6	A CT scan of the chest should be considered before a chest tube is inserted to treat a pneumothorax in PwCF, in the absence of an emergency.	GRADE D**	4	9 [2-9] UNCERTAIN (No Consensus)

**RECOMMENDATION OF LOW TO MODERATE CERTAINTY.

2.5. Atelectasis

The causes of atelectasis in CF are multifactorial and include: 1) mucus plugging due to impaired ion transport and mucociliary clearance [2,80–82]) chronic infection and inflammation, leading to increased mucus production and viscosity [3,81–83]) ABPA [4,84]) and congenital airway structural abnormalities [82]. These mechanisms often interact, creating a cycle of obstruction, infection, and inflammation that underlies the development of atelectasis in CF (see Fig. 7).

CT remains the most sensitive and specific imaging modality for diagnosing atelectasis in CF, but MRI offers a robust, radiation-free

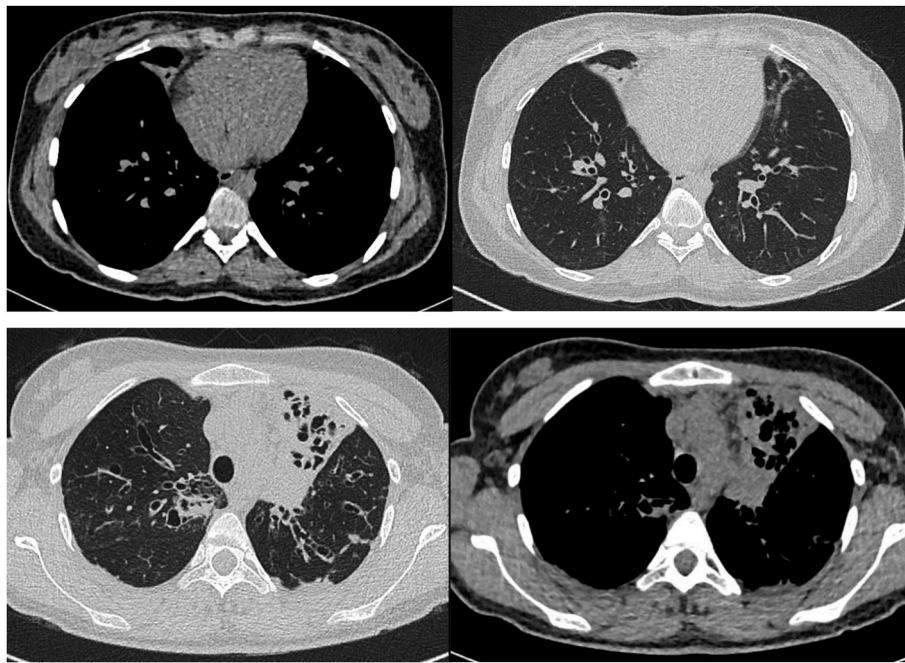


Fig. 7. Chronic atelectasis in the middle (upper panels) and left upper lobe (lower panels) shown axially (1 mm) in both lung and soft tissue windows (Courtesy Dr Marcel Opitz).

alternative with high diagnostic yield. Lung ultrasound is a useful adjunct for detecting subpleural atelectasis, particularly in children, while Multiple Breath Washout (MBW) provides complementary functional information but lacks anatomical specificity [85–88]. The choice of modality should be individualized based on patient age, clinical scenario, and the need to balance diagnostic accuracy with radiation safety.

Several papers have addressed the use of chest CT in evaluating atelectasis in CF [52]. A recent study specifically examined the prognostic significance of pulmonary atelectasis in CF, finding that persistent atelectasis is associated with worse lung function, increased exacerbations, and a higher frequency of comorbidities such as ABPA. The study also noted that bronchoscopy did not lead to clinico-radiological improvement in these patients, highlighting the importance of early detection and prevention of persistent atelectasis [84].

In our survey, the performance of a CT scan of the chest in the case of atelectasis in PwCF was considered useful by almost all respondents, 100% of adult physicians finding it useful, with 83% of paediatricians finding it useful, 12% not so useful and 5% not useful at all.

In summary, chest CT is the reference standard for detecting and monitoring atelectasis in CF, and its use is supported by both clinical studies and guidelines from the CFF.

3. Conclusion

Chest CT is justified in the presence of clinically significant haemoptysis, ABPA and multiple NTM positive cultures. However, no consensus was reached on use of CT in case of an atelectasis, or before the insertion of a chest tube in the case of pneumothorax. Overall, the clinical benefit of a chest CT scan in the above-mentioned clinical scenarios is variable and age dependent. Contrast CT is usually not required; the only need for contrast CT is haemoptysis. Improvements in therapeutic management (CFTR modulator and gene therapies) and the use of emerging CT technologies such as photon-counting CT (PCCT) enable substantial radiation dose reductions compared with conventional systems, with studies demonstrating reductions of approximately 45–66% while maintaining diagnostic image quality. In ultra-low-dose chest protocols, reductions of up to 65–90% have been reported,

approaching dose levels comparable to chest radiography in selected applications. These advances are driven by improved detector efficiency, reduced electronic noise, and enhanced image reconstruction techniques [89,90].

Consideration of other non-radiating imaging techniques such as lung MRI is an increasingly valuable adjunct to CT in assessing NTM lung disease in CF, particularly due to its lack of ionising radiation, enabling safer longitudinal monitoring in younger patients. MRI can also provide functional information, including ventilation and perfusion abnormalities, which may enhance detection of early or treatment-related changes. However, compared with CT, MRI has lower spatial resolution and reduced sensitivity for small nodules, early bronchiectasis and cavitation, key features in NTM disease. Consequently, CT remains the gold standard for diagnosis and structural characterisation, while MRI is best positioned as a complementary tool for serial monitoring and functional assessment [19,91,92].

CRedit authorship contribution statement

Michael Fayon: Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Project administration, Supervision, Validation, Visualization, Writing – original draft, Writing – review & editing. **Kate Hill:** Conceptualization, Data curation, Formal analysis, Methodology, Project administration, Supervision, Validation, Visualization, Writing – original draft, Writing – review & editing. **Luca Riberi:** Formal analysis, Methodology, Writing – original draft, Writing – review & editing. **Gael Dournes:** Formal analysis, Methodology, Writing – original draft, Writing – review & editing. **Pierre-Régis Burgel:** Formal analysis, Methodology, Writing – original draft, Writing – review & editing. **Barbara Messori:** Formal analysis, Methodology, Writing – original draft, Writing – review & editing. **Michael Waldron:** Methodology, Writing – original draft, Writing – review & editing. **Marcus Svedberg:** Formal analysis, Methodology, Writing – original draft, Writing – review & editing. **Elise Lammertyn:** Writing – original draft, Writing – review & editing. **Stojka Fustik:** Formal analysis, Methodology, Writing – original draft, Writing – review & editing. **Andrea Gramegna:** Formal analysis, Methodology, Writing – original draft, Writing – review & editing. **Mirjam Stahl:** Formal analysis, Methodology,

Writing – original draft, Writing – review & editing. **Anna Kerpel-Fronius:** Methodology, Writing – original draft, Writing – review & editing. **Maurizio Balbi:** Methodology, Writing – original draft, Writing – review & editing. **Pierluigi Ciet:** Conceptualization, Formal analysis, Methodology, Writing – original draft, Writing – review & editing. **Guillaume Chassagnon:** Methodology, Writing – original draft, Writing – review & editing. **Cinzia Ferrero:** Methodology, Writing – original draft, Writing – review & editing. **Sivagurunathan Sutharsan:** Formal analysis, Methodology, Writing – original draft, Writing – review & editing. **Marcel Opitz:** Formal analysis, Methodology, Writing – original draft, Writing – review & editing. **Eleni-Rosalina Andrinopoulou:** Formal analysis, Methodology, Writing – original draft, Writing – review & editing. **Michael Maher:** Conceptualization, Formal analysis, Methodology, Writing – original draft, Writing – review & editing. **Jamie Duckers:** Methodology, Writing – original draft, Writing – review & editing. **Harm Tiddens:** Conceptualization, Formal analysis, Methodology, Writing – original draft, Writing – review & editing. **Isabelle Sermet-Gaudelus:** Conceptualization, Formal analysis, Methodology, Writing – original draft, Writing – review & editing.

Declaration of competing interest

The authors declare the following financial interests/personal relationships which may be considered as potential competing interests:

M Fayon has a patient in the field of neonatal lung diseases: patent #FR3105004 & is President of the Pulmonology Society of the Indian Ocean.

K Hill has received support for attending meetings from ECFS-CTN, is a non-voting member of the ECFS-CTN Executive Committee and paid employee of the ECFS-CTN.

P-R Burel has received institutional support from GSK, Vertex, & consulting fees from Asta-Zeneca, Chiesi, GSK, Insmmed, MSD, Vertex, Viatrix and Zambon; support for attending meetings from Asta-Zeneca & Chiesi.

Elise Lammetyn has received institutional support from Patient Centrics & Roche BeLux & is a member of the ECFS Patient Registry Scientific Committee.

A Gramegna has received payment or honoraria from Vertex and Chiesi; support for attending meetings from Foodar, Neupharma, Menarini & is ERS Group 10.3 Chair (Adult CF).

M Stahl has received institutional support from the German Research Foundation (Deutsche Forschungsgemeinschaft, DFG); grants or contracts from Vertex Pharmaceuticals Independent RIA grant & for various clinical studies; payment or honoraria from Vertex Pharmaceuticals: honoraria for presentations including LEAD meetings; participation on DSMB for Vertex Pharmaceuticals: Advisory Board & is Chairman FGM, Secretary Group CF ERS & Treasurer GPP.

A-K Fronius is the President elect of the Hungarian Society of Radiologists & Member of the Board.

P Ciet has received speaker fees from Vertex, Siemens Healthineers; grants or contracts from the Research Dutch Council, AIC Horizon Pathfinder and General Electric Healthcare; payment or honoraria for lectures from Vertex, Siemens Healthineers & is Chair of the Cardiothoracic Taskforce ESPR & Member of the Research Commission ESPR.

G Chassagnon has received consulting fees from Gleamer SA, and payment or honoraria from Cheisi and Boehringer Ingelheim.

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J Duckers has received payment or honoraria from Chiesi Pharmaceuticals, Insmmed, Pfizer & Vertex & is a member of DSMB Pharmaceuticals & Chair British Thoracic Society Specialist advisory group, Chair UK CF Registry.

H Tiddens, Erasmus MC has received support from NIH, CFF, Sophia Foundation & NHMRC. The Erasmus MC holds a license agreement with Thirona and Resonance Health for the use of PRAGMA-CF. He has

advised on Image analysis of dataset by Erasmus MC-Lung Analysis and Thirona & received payment or honoraria from Vertex for faculty for ADVANCE course, faculty for webinar on imaging. Erasmus MC and Thirona reimburse costs made for participating in relevant conferences as part of his work. He is Chair of the Imaging Standardization Committee ECFS-CTN & Emeritus Professor Erasmus MC; Founder, and former director Erasmus MC Lung Analysis Core laboratory & CMO Thirona.

I Sermet has received grants or contracts from the CFF grant to support paediatric research; has received institutional funding from Vertex Pharmaceuticals for academic research projects and is an investigator in Vertex Pharmaceuticals trials & consulting fees from Vertex and Eloxx for participation on Scientific Advisory Board. She is ECFS-CTN Standardisation Committee Chair & previous ECFS Board member & has received non-financial support for PTC Therapeutics for participation in Scientific Discussions.

M Maher, M Opitz, C Ferrero, M Balbi, S Fustik, M Svedberg, B Messori, M Waldron, G Dournes, E-R Andrinopoulou & L Riberi have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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