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Clinical indications and scanning protocols for Chest CT in children with cystic fibrosis: a survey of UK tertiary centres

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Clinical indications and scanning protocols for Chest CT in children with cystic fibrosis: a survey of UK tertiary centres

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Data Sharing Statement: All data from this article is available upon request to the corresponding author.

Abstract

Objectives

Chest computerised tomography (CT) is increasingly used to monitor disease progression in children with cystic fibrosis (CF) but there is no national guideline regarding its use. Our objective was to assess the indications for undertaking chest CT and the protocols used to obtain scans.

Design, Setting and participants

An electronic questionnaire was developed to assess clinicians views on chest CT in children with CF. It included general questions on perceived benefits and specific questions about its role in five clinical scenarios. It was sent to the clinical lead in 27 UK paediatric CF Centres. A separate questionnaire was developed to collect the technical details of chest CT in children with CF. It was sent to the superintendent radiographer at each of the 27 centres.

Results:

Responses were obtained from 27 (100%) clinical leads and 22 (81%) superintendent radiographers. 93% clinicians reported chest CT useful in monitoring disease progression and 70% said it frequently altered management. Only 5 (19%) undertook routine scans. To aid diagnosis, 81% performed chest CT in non-tuberculous mycobacterial disease and 15% in allergic bronchopulmonary aspergillosis. There was wide variation in the perceived need for and/or timing of chest CT in children with reduced lung function with no benefit from intravenous antibiotics, new cystic changes on chest x-ray, and lobar collapse. The radiographers reported using a mixture of helical (volumetric) and axial scans depending on the clinical question, the age and the co-operation of the child. When indicated, 6 (27%) used sedation and 16 (73%) general anaesthetic. Only 1 (5%) used intravenous contrast routinely and 3 (14%) obtained expiratory images routinely.

Conclusions:

There is marked variation in the use of chest CT in children with CF and in the scan protocols. The lack of a national guideline is likely to be contributing to this lack of standardisation.

What is known about the subject?

1. A computerised tomography (CT) scan is the gold standard imaging modality for assessment of structural lung disease in cystic fibrosis (CF).
2. The use of chest CT is increasing and in some European Paediatric CF centres, scans are routinely performed biennially
3. There is no UK national guideline for the use of chest CT in CF

What this study adds

1. There was marked variation amongst UK centres in the clinical indications for chest CT in children with CF
2. There was marked variation in the protocols used by radiographers when obtaining chest CT scans in children with CF
3. These differences highlight the need for a national guideline

Introduction

Cystic fibrosis (CF) lung disease is characterised by lower airway infection and chronic inflammation leading to lung damage and progressive respiratory failure.[1] Accurate assessment of lung disease in children with CF is vital for monitoring disease progression and guiding treatment.[2] Computerised tomography (CT) is the gold standard for assessing the structural component of CF lung disease.[3] It is sensitive enough to detect early bronchiectasis and gas trapping in infants diagnosed by newborn screening[4,5] and in older children and adults, can detect changes before they become apparent on pulmonary function testing.[6] This has led to increased use of chest CT in children with CF and in some European centres, routine scans are performed biennially.[7,8] It is also accepted as a useful outcome measure in CF clinical trials[9] although this is limited by poor inter and intra-observer agreement for the scoring systems, especially in young children.[10] The benefits

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3 of chest CT must be balanced against the subsequent lifetime risk of malignancy associated with
4 ionising radiation.[7,11] This is relevant in CF as life expectancy has increased beyond the age at
5 which such malignancies present.[12] The introduction of modern scanners and the use of paediatric
6 specific scan protocols has reduced the radiation dose associated with CT scans.[13] Despite this, the
7 cumulative radiation dose for children with CF is substantial and chest CT is the major
8 contributor.[14,15]

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11 The role for chest CT is defined in diagnostic guidelines for CF complications such as allergic
12 bronchopulmonary aspergillosis (ABPA) and non-tuberculous mycobacterium (NTM) disease[16,17]
13 but there is no clear guidance about its general use in children with CF. As the first step toward
14 developing a guideline, we assessed current practice regarding clinicians views on the indications for
15 scanning and the protocols used by radiologist / radiographers.

16 17 18 19 20 21 22 23 24 25 26 27 28 **Aims**

29
30 To assess current UK practice regarding the indications for undertaking chest CT in children with CF
31 and the protocols used for performing these scans.

32 33 34 35 36 37 38 39 40 41 42 43 44 45 46 47 48 49 50 51 52 53 54 55 56 57 58 59 60 **Methods**

A questionnaire was developed to assess the views of clinicians on chest CT in children with CF. It collected information on the perceived benefit of chest CT in monitoring disease progression, the likelihood of the scan altering management, the use of baseline scans, knowledge of the associated radiation dose and discussion of this with the parent / guardian. The questionnaire also contained five case vignettes which assessed if and when a chest CT would be undertaken in a child with NTM, with reduced lung function and no improvement with intravenous (IV) antibiotics, with new CXR changes, with ABPA and with lobar collapse. This questionnaire can be seen in Appendix 1A. An electronic link to this questionnaire was sent to the clinical lead at each of the 27 UK paediatric CF centres who were asked to respond on behalf of his/her centre.

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3 A separate questionnaire was developed to identify the technical details of chest CT when
4 performed in children with CF. It collected data on the make and model of scanner, the type of scans
5 performed, the use of sedation or general anaesthetic, the use of IV contrast and the acquisition of
6 expiratory images. This questionnaire can be seen in Appendix 1B. An electronic link to this
7 questionnaire was sent to the superintendent radiographer at each of the 27 UK paediatric CF
8 centres.
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16 **Results**

17 Clinical Indications

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19 Responses were obtained from all 27 Clinical Leads. Chest CT was thought to be useful in monitoring
20 disease progression by 25/27 (93%) and frequently alter management by 19/27 (70%). Only 5/27
21 (19%) centres undertake a baseline chest CT in an otherwise well child. In these centres, the mean
22 (SD) age for acquiring a baseline scan was 8 (4.3) years. Three of those five centres continue to
23 perform surveillance scans every 4 (1.4) years. 24/27 (89%) reported being aware of the radiation
24 dose associated with chest CT at their centre. The reported dose varied from the equivalent of two
25 CXRs (0.04 mSv) to 2.1mSv (equivalent to approximately 102 CXRs). Discussion of the potential
26 harmful effects of chest CT was reported as taking place 'often' or 'always' by 20/27 (74%). A
27 summary of the responses regarding the need for and timing of chest CT in five common scenarios is
28 reported in Table 1. There was a low level of overall agreement. Only five (19%) clinicians managed
29 all five case scenarios in the same way. The remaining 22 respondents each gave a unique
30 combination of answers.
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Table 1: Summary of responses regarding the 'need for' or 'timing of' chest CT in five common scenarios.

		Responses	
		Number	%
NTM Pulmonary Disease	Yes	22	81%
	No	5	19%
Reduced FEV₁ with no response to intravenous antibiotics	Yes - At the same time as bronchoscopy	6	22%
	Yes - If bronchoscopy does not reveal cause	19	70%
	Not at any point	2	7%
New cystic changes on CXR	Yes	15	56%
	No	12	44%
Allergic bronchopulmonary aspergillosis	Yes	4	15%
	No	23	85%
Lobar collapse	Yes - As soon as CXR shows lobar collapse	1	4%
	Yes - If IV antibiotics and physiotherapy unsuccessful at re-inflating lobe	5	19%
	Yes - If bronchoscopy unsuccessful at re-inflating lobe	15	56%
	Not at any point	6	22%

NTM: non-tuberculous mycobacteria, FEV₁: forced expiratory volume in one second, CXR: chest x-ray.

Radiological Protocols

Responses were obtained from the superintendent radiographer at 22/27 (81%) centres. Fourteen different types of scanners were used across these centres. When performing chest CT in children with CF, a mixture of helical and axial scans were used. The decision on the type of scan was made by the radiologist based on the clinical question, the age of the child and the ability of the child to co-operate. Only 6 (27%) centres used sedation. Indications for sedation included the child being uncooperative, the child having learning difficulties or a previous failed CT without sedation. General anaesthetic (GA) was used by 16 (73%) of centres. Indications for GA included the child being unable to co-operate with a breath hold, being of a young age or having learning difficulties. Only one (5%) centre reported the routine use of contrast and three (14%) routinely obtained expiratory images. When expiratory images were obtained, 14 (64%) used breath-holding command, five (23%) relied on ventilation by the anaesthetist during GA and three (14%) used decubitus positioning.

Discussion

To our knowledge this is the first time UK practice regarding the clinical indications and protocols used for chest CT in children with CF has been analysed. We have identified marked variation on the clinical reason for undertaking the scan and the protocol used to acquire it. This highlights the need for a national guideline to standardise and promote best practice.

The responses from clinical leads confirmed chest CT scans are perceived as a useful tool for monitoring the progression of CF lung disease and does influence clinical management. Despite this, less than a fifth of centres undertake a baseline scan and no UK centre is performing routine biennial scans as practiced in some parts of Europe.[15] The clinical vignettes demonstrated good levels of agreement that chest CT was needed for the diagnosis of NTM disease but not for the diagnosis of ABPA. This reflects the advice in the relevant guidelines.[16,17] In contrast, there was wide variation in the use of chest CT in other common CF clinical scenarios (reduced FEV1 with no response to IV antibiotics, new cystic changes on CXR and lobar collapse) for which there are currently no guidelines. The difference in the use and timing of chest CT at UK paediatric CF centres will have an influence in the cumulative radiation exposure and lifetime cancer risk for children with CF at these centres.

The benefits of chest CT must be balanced against the increased cancer risk associated with cumulative exposure to ionising radiation.[7,11] This is particularly important in CF as affected individuals undergo repeated radiological investigations and show increased incidence of certain digestive tract malignancies.[12] The use of protocols specific to patient size and the region scanned has reduced the radiation dose associated with CT scans.[13] Despite this, the cumulative radiation exposure in children with CF is substantial with chest CT being the biggest contributor.[14] A computational model which calculated excess mortality in a CF cohort associated with radiation from annual or biennial chest CT showed that routine lifelong CT scans carry a low risk of radiation-induced mortality.[15] This is despite the cumulative radiation exposure in an 18 year old with CF

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3 from chest CT alone being approximately 9mSv if biennial scans have been performed and 18mSv if
4 annual scans are performed. This compares to 2.8mSv when chest CT is only performed when
5 clinically indicated.[14] To put these doses into context, the annual background radiation dose in the
6 UK is approximately 2.7mSv.[18] The radiation dose associated with a CT scan depends on the region
7 of the body being scanned, the type of scan, the age/size of the child and dose saving features used.
8 It is therefore unsurprising that although clinicians reported being aware of the radiation dose
9 associated with chest CT at their centre, the reported dose varied more than 500 fold (0.04 to 2.1
10 mSv). This difference is far higher than can be explained by the different protocols. A recent single
11 centre study reported the Estimated Effective Dose from chest CT in children to be 0.57-2.79 mSv for
12 helical scans and 0.22-0.59 mSv for axial scans.[14]

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Most young children are unable to cooperate with voluntary breath hold instructions thereby
necessitating sedation or GA for lung volume standardisation and the acquisition of high quality
images. Modern flash scanners can perform a chest CT in 1-2 seconds which allows scans to be
performed during free tidal-volume breathing. This reduces the need for sedation or GA. We are not
aware of any studies comparing the sensitivity of tidal-volume breathing scans and pressure-
controlled scans at detecting structural lung changes. Of the centres that responded to our survey,
GA was used more frequently than sedation (73% vs 27%). This may relate to the risks of sedation
being unsuccessful / inadequate or causing hypoxia. In a large prospective study these were
reported as 23% and 2.9% respectively.[19] There is a risk of iatrogenic atelectasis on chest CT
performed under GA.[20] In children with CF it can therefore be difficult to determine if atelectasis
seen on a scan is caused by the GA or the underlying CF lung disease. The risk of atelectasis can be
reduced by the use of lung recruitment manoeuvres and controlled-ventilation.[20]

We accept there are limitations to this survey. Although responses were obtained from the clinical
lead at each UK centre, there may be variation in practice between the consultants at each centre.
We purposefully kept the case vignettes brief to maximise the response rate and minimise confusion

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3 but this potentially means they did not accurately reflect clinical practice. Despite multiple attempts,
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5 we were unable to obtain a response from the superintendent radiographer at five centres.
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8 **Conclusions**

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10 We have identified marked variation in the use of chest CT scans in children with CF and differences in
11
12 the protocols used when undertaking these scans. Guidance on the indications for Chest CT in
13
14 children with CF and recommendations on protocols to optimise image quality and limit radiation
15
16 exposure would be helpful at a national level. The choice of protocol, however, is also dependent on
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18 the clinician providing enough clinical information so it is clear what question the scan is trying to
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20 answer.
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CT in CF - Clinicians

Which cystic fibrosis centre do you work at?

Q1 Do you think CT is a useful tool in monitoring disease progression in children with cystic fibrosis?

- Yes
 No

Q1 Comments

Q2 Is it your impression that CT scan results frequently alter the management in children with cystic fibrosis?

- Yes
 No

Q2 Comments

Q3a In a clinically well child, would you undertake a baseline CT scan?

- Yes
 No

Q3a Comments

Answer If In a clinically well child, would you undertake a baseline CT scan at some point?

Yes Is Selected

Q3b At what age would you request this scan?

_____ Age (years)

Answer If In a clinically well child, would you undertake a baseline CT scan at some point?

Yes Is Selected

Q4a Would you repeat the CT scan after a period of time, even if the child remains well?

- Yes
 No

Answer If If the child remains well, would you undertake a repeat CT scan at some point?

Yes Is Selected

Q4b How many years after the baseline CT scan would you do this?

_____ Years

Q5 Are you aware of the radiation dose that is associated with a CT scan at your centre?

- Yes
 No

Q5 Comments

Q6 Are the potential harmful effects of CT scans discussed with the children and parents?

- Never
- Rarely
- Sometimes
- Often
- Always

Q6 com If so, what information do you provide?

Intro 2 The final 5 questions concern the use of CT scans in different clinical scenarios.

Sc1 A 10 year old boy has isolated Mycobacterium abscessus on 3 consecutive sputum samples. His FEV1 has also reduced from 95% to 80% predicted. Would you perform a CT scan to aid in the diagnosis of non-tuberculous mycobacterium disease?

- Yes
- No

Sc1 Comments

Sc2 A 10 year old girl with CF has deteriorated over a 6 month period. She has developed a chronic wet cough and her FEV1 has reduced from 100% predicted to 80%. Cough swabs have only isolated mixed flora and a chest x-ray did not show any new changes. A course of intravenous antibiotics had little effect and so a flexible bronchoscopy was requested. Would you order a CT scan for this girl?

- No - not at any point
- Yes - request at the same time as bronchoscopy
- Yes - but only if the bronchoscopy failed to identify a cause for the deterioration

Sc2 Comments

Sc3a A 10 year old boy with CF is noted to have increased bronchovascular markings and new cystic changes on his annual review chest x-ray. He denies any new symptoms and his spirometry is stable (FEV1 85%). Would you perform a CT scan in this boy?

- Yes
- No

Answer If 'A 10 year old boy with CF is noted to have increased bronchovascular markings and new cystic cha...' Yes Is Selected

Sc3b Is there a minimum time since his last CT scan that you would want to see elapsed before you repeat the CT scan?

- Yes
- No

Answer If 'Is there a minimum time since his last CT scan that you would want to see elapsed before you repeat the CT scan?' Yes Is Selected

Sc3c What is this minimum time in months?

Sc3 Comments

Sc4 A 10 year old girl with CF has deteriorated over a 6 month period. She has increased cough and her FEV1 has fallen from 85% to 70% predicted. There is limited improvement with course of intravenous antibiotics. Her total IgE is 1500 and she has positive specific IgE and precipitins to *Aspergillus fumigatus*. There are no new changes on her CXR. Would you routinely perform a CT scan to aid the diagnosis of ABPA?

- Yes
- No

Sc4 Comments

Sc5 A 10 year old girl with CF develops a left lower lobe collapse during an infective exacerbation. Despite admission for 2 weeks intravenous antibiotics and intensive physiotherapy, the lobe does not re-inflate. She is therefore booked for a flexible bronchoscopy. Would you undertake a CT scan in this girl?

- No - not at any point
- Yes - as soon as the chest x-ray confirmed the left lower lobe collapse
- Yes - but only after two weeks of antibiotics and physiotherapy have failed to re-inflate the lobe
- Yes - but only if bronchoscopy is unsuccessful at re-inflating the lobe

Sc5 Comments

Final Do you have any further comments regarding the use of CT scans in children with cystic fibrosis?

CT Radiologists

Q1 Which centre are you from?

Q2 What is the make and model of the primary CT scanner that is used to scan children at your centre?

- Siemens
- GE
- Toshiba
- Phillips
- Other (please specify) _____

Q3 Model:

Q4 What type of chest CT scans do you perform in children with CF? (Select all applicable)

- Volume / helical scan
- HRCT / axial

Q5 What influences your choice of protocol (i.e. helical vs. axial) e.g. age of patient, patient co-operation etc...?

Q6 When performing chest CT, do you routinely acquire expiratory images in children with CF?

- Yes
- No
- Don't know

Q7 If you do acquire expiratory images (even if non-routine), how is this done?

- Breath holding
- Decubitus positioning
- Other (please specify) _____
- Never acquire
- Don't know

Q8 Do you routinely use contrast when performing chest CT scans in children with CF?

- Yes
- No
- Don't know

Answer If Do you routinely use contrast? No Is Selected

Q8A Are there any circumstances in which you would use contrast to scan the chest of a child with CF? Please give details

1
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3 Q9 Do you ever scan children with CF under general anaesthetic?

- 4 Yes
5 No
6 Don't know
7

8 Answer If Do you ever scan children with CF under general anaesthetic? Yes Is Selected

9 Q9A Under what circumstances would you use general anaesthetic (e.g. in which age
10 groups)?
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13 Q10 Do you ever use sedation to scan children with CF?

- 14 Yes
15 No
16 Don't know
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18 Answer If Do you ever use sedation to scan children with CF. Yes Is Selected

19 Q10A Under what circumstances would you use sedation (e.g. in which age groups)?
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22 Q11 Do you have any further comments regarding the use of CT scans in children with CF?
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Abstract

Objectives

Chest computerised tomography (CT) is increasingly used to monitor disease progression in children with cystic fibrosis (CF) but there is no national guideline regarding its use. Our objective was to assess the indications for undertaking chest CT and the protocols used to obtain scans.

Design, Setting and participants

An electronic questionnaire was developed to assess clinicians views on chest CT in children with CF. It included general questions on perceived benefits and specific questions about its role in five clinical scenarios. It was sent to the clinical lead in 27 UK paediatric CF Centres. A separate questionnaire was developed to collect the technical details of chest CT in children with CF. It was sent to the superintendent radiographer at each of the 27 centres.

Results:

Responses were obtained from 27 (100%) clinical leads and 22 (81%) superintendent radiographers. 93% clinicians reported chest CT useful in monitoring disease progression and 70% said it frequently altered management. Only 5 (19%) undertook routine scans. To aid diagnosis, 81% performed chest CT in non-tuberculous mycobacterial disease and 15% in allergic bronchopulmonary aspergillosis. There was wide variation in the perceived need for and/or timing of chest CT in children with reduced lung function with no benefit from intravenous antibiotics, new cystic changes on chest x-ray, and lobar collapse. The radiographers reported using a mixture of helical (volumetric) and axial scans depending on the clinical question, the age and the co-operation of the child. When indicated, 6 (27%) used sedation and 16 (73%) general anaesthetic. Only 1 (5%) used intravenous contrast routinely and 3 (14%) obtained expiratory images routinely.

Conclusions:

There is marked variation in the use of chest CT in children with CF and in the scan protocols. The lack of a national guideline is likely to be contributing to this lack of standardisation.

What is known about the subject?

1. A computerised tomography (CT) scan is the gold standard imaging modality for assessment of structural lung disease in cystic fibrosis (CF).
2. The use of chest CT is increasing and in some European Paediatric CF centres, scans are routinely performed biennially
3. There is no UK national guideline for the use of chest CT in CF

What this study adds

1. There was marked variation amongst UK centres in the clinical indications for chest CT in children with CF
2. There was marked variation in the protocols used by radiographers when obtaining chest CT scans in children with CF
3. These differences highlight the need for a national guideline

Introduction

Cystic fibrosis (CF) lung disease is characterised by lower airway infection and chronic inflammation leading to lung damage and progressive respiratory failure.[1] Accurate assessment of lung disease in children with CF is vital for monitoring disease progression and guiding treatment.[2] Computerised tomography (CT) is the gold standard for assessing the structural component of CF lung disease.[3] It is sensitive enough to detect early bronchiectasis and gas trapping in infants diagnosed by newborn screening[4,5] and in older children and adults, can detect changes before they become apparent on pulmonary function testing.[6] This has led to increased use of chest CT in children with CF and in some European centres, routine scans are performed biennially.[7,8] It is also accepted as a useful outcome measure in CF clinical trials[9] although this is limited by poor inter and intra-observer agreement for the scoring systems, especially in young children.[10] The benefits

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3 of chest CT must be balanced against the subsequent lifetime risk of malignancy associated with
4 ionising radiation.[7,11] This is relevant in CF as life expectancy has increased beyond the age at
5 which such malignancies present.[12] The introduction of modern scanners and the use of paediatric
6 specific scan protocols has reduced the radiation dose associated with CT scans.[13] Despite this, the
7 cumulative radiation dose for children with CF is substantial and chest CT is the major
8 contributor.[14,15]

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11 The role for chest CT is defined in diagnostic guidelines for CF complications such as allergic
12 bronchopulmonary aspergillosis (ABPA) and non-tuberculous mycobacterium (NTM) disease[16,17]
13 but there is no clear guidance about its general use in children with CF. As the first step toward
14 developing a guideline, we assessed current practice regarding clinicians views on the indications for
15 scanning and the protocols used by radiologist / radiographers.

16 17 18 19 20 21 22 23 24 25 26 27 28 **Aims**

29
30 To assess current UK practice regarding the indications for undertaking chest CT in children with CF
31 and the protocols used for performing these scans.

32 33 34 35 36 37 38 39 40 41 42 43 44 45 46 47 48 49 50 51 52 53 54 55 56 57 58 59 60 **Methods**

A questionnaire was developed to assess the views of clinicians on chest CT in children with CF. It collected information on the perceived benefit of chest CT in monitoring disease progression, the likelihood of the scan altering management, the use of baseline scans, knowledge of the associated radiation dose and discussion of this with the parent / guardian. The questionnaire also contained five case vignettes which assessed if and when a chest CT would be undertaken in a child with NTM, with reduced lung function and no improvement with intravenous (IV) antibiotics, with new chest x-ray (CXR) changes, with ABPA and with lobar collapse. This questionnaire can be seen in Appendix 1A. An electronic link to this questionnaire was sent to the clinical lead at each of the 27 UK paediatric CF centres who were asked to respond on behalf of his/her centre.

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3 A separate questionnaire was developed to identify the technical details of chest CT when
4 performed in children with CF. It collected data on the make and model of scanner, the type of scans
5 performed, the use of sedation or general anaesthetic, the use of IV contrast and the acquisition of
6 expiratory images. This questionnaire can be seen in Appendix 1B. An electronic link to this
7 questionnaire was sent to the superintendent radiographer at each of the 27 UK paediatric CF
8 centres.
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16 **Results**

17 Clinical Indications

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19 Responses were obtained from all 27 Clinical Leads. Chest CT was thought to be useful in monitoring
20 disease progression by 25/27 (93%) and frequently alter management by 19/27 (70%). Only 5/27
21 (19%) centres undertake a baseline chest CT in an otherwise well child. In these centres, the mean
22 (SD) age for acquiring a baseline scan was 8 (4.3) years. Three of those five centres continue to
23 perform surveillance scans every 4 (1.4) years. 24/27 (89%) reported being aware of the radiation
24 dose associated with chest CT at their centre. The reported dose varied from the equivalent of two
25 CXRs (0.04 mSv) to 2.1mSv (equivalent to approximately 102 CXRs). Discussion of the potential
26 harmful effects of chest CT was reported as taking place 'often' or 'always' by 20/27 (74%). A
27 summary of the responses regarding the need for and timing of chest CT in five common scenarios is
28 reported in Table 1. There was a low level of overall agreement. Only five (19%) clinicians managed
29 all five case scenarios in the same way. The remaining 22 respondents each gave a unique
30 combination of answers.
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Table 1: Summary of responses regarding the 'need for' or 'timing of' chest CT in five common scenarios.

		Responses	
		Number	%
NTM Pulmonary Disease	Yes	22	81%
	No	5	19%
Reduced FEV₁ with no response to intravenous antibiotics	Yes - At the same time as bronchoscopy	6	22%
	Yes - If bronchoscopy does not reveal cause	19	70%
	Not at any point	2	7%
New cystic changes on CXR	Yes	15	56%
	No	12	44%
Allergic bronchopulmonary aspergillosis	Yes	4	15%
	No	23	85%
Lobar collapse	Yes - As soon as CXR shows lobar collapse	1	4%
	Yes - If IV antibiotics and physiotherapy unsuccessful at re-inflating lobe	5	19%
	Yes - If bronchoscopy unsuccessful at re-inflating lobe	15	56%
	Not at any point	6	22%

NTM: non-tuberculous mycobacteria, FEV₁: forced expiratory volume in one second, CXR: chest x-ray.

Radiological Protocols

Responses were obtained from the superintendent radiographer at 22/27 (81%) centres. Fourteen different types of scanners were used across these centres. When performing chest CT in children with CF, a mixture of helical and axial scans were used. The decision on the type of scan was made by the radiologist based on the clinical question, the age of the child and the ability of the child to co-operate. Only 6 (27%) centres reported using sedation. Indications for sedation included the child being uncooperative, the child having learning difficulties or a previous failed CT without sedation. General anaesthetic (GA) was used if necessary by 16 (73%) of centres. Indications for GA included the child being unable to co-operate with a breath hold, being of a young age or having learning difficulties. Only one (5%) centre reported the routine use of contrast and three (14%) routinely obtained expiratory images. When expiratory images were obtained, 14 (64%) used breath-holding command, five (23%) relied on ventilation by the anaesthetist during GA and three (14%) used decubitus positioning.

Discussion

To our knowledge this is the first time UK practice regarding the clinical indications and protocols used for chest CT in children with CF has been analysed. We have identified marked variation on the clinical reason for undertaking the scan and the protocol used to acquire it. This highlights the need for a national guideline to standardise and promote best practice.

The responses from clinical leads confirmed chest CT scans are perceived as a useful tool for monitoring the progression of CF lung disease and does influence clinical management. Despite this, less than a fifth of centres undertake a baseline scan and no UK centre is performing routine biennial scans as practiced in some parts of Europe.[15] The clinical vignettes demonstrated good levels of agreement that chest CT was needed for the diagnosis of NTM disease but not for the diagnosis of ABPA. This reflects the advice in the relevant guidelines.[16,17] In contrast, there was wide variation in the use of chest CT in other common CF clinical scenarios (reduced FEV1 with no response to IV antibiotics, new cystic changes on CXR and lobar collapse) for which there is currently no UK guideline. The National Institute of Clinical Excellence (NICE) document: Cystic fibrosis: diagnosis and management, suggests clinicians 'consider a low-dose chest CT scan for children with cystic fibrosis who have not had a chest CT scan before, to detect features that other tests (such as CXR) would miss (for example early bronchiectasis).'[18] Whilst this is useful it does not give specific advice about when chest CT should and should not be performed. The CF Foundation recommends against the use chest CT scans for routine surveillance in children under 2 years.[19] In older children it recommends consideration of chest CT as an alternative to CXR to monitor progression of lung disease but no specific advice is given.[20] The difference in the use and timing of chest CT at UK paediatric CF centres will have an influence in the cumulative radiation exposure and lifetime cancer risk for children with CF at these centres.

The benefits of chest CT must be balanced against the increased cancer risk associated with cumulative exposure to ionising radiation.[7,11] This is particularly important in CF as affected

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3 individuals undergo repeated radiological investigations and show increased incidence of certain
4 digestive tract malignancies.[12] The use of protocols specific to patient size and the region scanned
5 has reduced the radiation dose associated with CT scans.[13] Despite this, the cumulative radiation
6 exposure in children with CF is substantial with chest CT being the biggest contributor.[14] A
7 computational model which calculated excess mortality in a CF cohort associated with radiation
8 from annual or biennial chest CT showed that routine lifelong CT scans carry a low risk of radiation-
9 induced mortality.[15] This is despite the cumulative radiation exposure in an 18 year old with CF
10 from chest CT alone being approximately 9mSv if biennial scans have been performed and 18mSv if
11 annual scans are performed. This compares to 2.8mSv when chest CT is only performed when
12 clinically indicated.[14] To put these doses into context, the annual background radiation dose in the
13 UK is approximately 2.7mSv.[21] The radiation dose associated with a CT scan depends on the region
14 of the body being scanned, the type of scan, the age/size of the child and dose saving features used.
15 It is therefore unsurprising that although clinicians reported being aware of the radiation dose
16 associated with chest CT at their centre, the reported dose varied more than 500 fold (0.04 to 2.1
17 mSv). This difference is far higher than can be explained by the different protocols. A recent single
18 centre study reported the Estimated Effective Dose from chest CT in children to be 0.57-2.79 mSv for
19 helical scans and 0.22-0.59 mSv for axial scans.[14]

20
21 Most young children are unable to cooperate with voluntary breath hold instructions thereby
22 necessitating sedation or GA for lung volume standardisation and the acquisition of high quality
23 images. Modern flash scanners can perform a chest CT in 1-2 seconds which allows scans to be
24 performed during free tidal-volume breathing. This reduces the need for sedation or GA. We are not
25 aware of any studies comparing the sensitivity of tidal-volume breathing scans and pressure-
26 controlled scans at detecting structural lung changes. Of the centres that responded to our survey,
27 GA was used more frequently than sedation (73% vs 27%). This may relate to the risks of sedation
28 being unsuccessful / inadequate or causing hypoxia. In a large prospective study these were
29 reported as 23% and 2.9% respectively.[22] There is a risk of iatrogenic atelectasis on chest CT

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3 performed under GA.[23] In children with CF it can therefore be difficult to determine if atelectasis
4
5 seen on a scan is caused by the GA or the underlying CF lung disease. The risk of atelectasis can be
6
7 reduced by the use of lung recruitment manoeuvres and controlled-ventilation.[23]
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10 We accept there are limitations to this survey. Although responses were obtained from the clinical
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12 lead at each UK centre, there may be variation in practice between the consultants at each centre.
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14 We purposefully kept the case vignettes brief to maximise the response rate and minimise confusion
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16 but this potentially means they did not accurately reflect clinical practice. Despite multiple attempts,
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18 we were unable to obtain a response from the superintendent radiographer at five centres.
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21 **Conclusions**

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24 We have identified marked variation in the use of chest CT scans in children with CF and differences in
25
26 the protocols used when undertaking these scans. Guidance on the indications for Chest CT in
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28 children with CF and recommendations on protocols to optimise image quality and limit radiation
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30 exposure would be helpful at a national level. The choice of protocol, however, is also dependent on
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32 the clinician providing enough clinical information so it is clear what question the scan is trying to
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34 answer.
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CT in CF - Clinicians

Which cystic fibrosis centre do you work at?

Q1 Do you think CT is a useful tool in monitoring disease progression in children with cystic fibrosis?

- Yes
 No

Q1 Comments

Q2 Is it your impression that CT scan results frequently alter the management in children with cystic fibrosis?

- Yes
 No

Q2 Comments

Q3a In a clinically well child, would you undertake a baseline CT scan?

- Yes
 No

Q3a Comments

Answer If In a clinically well child, would you undertake a baseline CT scan at some point?
Yes Is Selected

Q3b At what age would you request this scan?

_____ Age (years)

Answer If In a clinically well child, would you undertake a baseline CT scan at some point?
Yes Is Selected

Q4a Would you repeat the CT scan after a period of time, even if the child remains well?

- Yes
 No

Answer If If the child remains well, would you undertake a repeat CT scan at some point?
Yes Is Selected

Q4b How many years after the baseline CT scan would you do this?

_____ Years

Q5 Are you aware of the radiation dose that is associated with a CT scan at your centre?

- Yes
 No

Q5 Comments

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2
3 Q6 Are the potential harmful effects of CT scans discussed with the children and parents?

- 4 Never
5 Rarely
6 Sometimes
7 Often
8 Always
9

10
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12 Q6 com If so, what information do you provide?

13
14 Intro 2 The final 5 questions concern the use of CT scans in different clinical scenarios.

15
16 Sc1 A 10 year old boy has isolated Mycobacterium abscessus on 3 consecutive sputum
17 samples. His FEV1 has also reduced from 95% to 80% predicted. Would you perform a CT
18 scan to aid in the diagnosis of non-tuberculous mycobacterium disease?

- 19 Yes
20 No
21
22

23
24 Sc1 Comments

25
26 Sc2 A 10 year old girl with CF has deteriorated over a 6 month period. She has developed a
27 chronic wet cough and her FEV1 has reduced from 100% predicted to 80%. Cough swabs
28 have only isolated mixed flora and a chest x-ray did not show any new changes. A course of
29 intravenous antibiotics had little effect and so a flexible bronchoscopy was
30 requested. Would you order a CT scan for this girl?

- 31 No - not at any point
32 Yes - request at the same time as bronchoscopy
33 Yes - but only if the bronchoscopy failed to identify a cause for the deterioration
34
35

36
37 Sc2 Comments

38
39 Sc3a A 10 year old boy with CF is noted to have increased bronchovascular markings and
40 new cystic changes on his annual review chest x-ray. He denies any new symptoms and his
41 spirometry is stable (FEV1 85%). Would you perform a CT scan in this boy?

- 42 Yes
43 No
44
45

46
47 Answer If 'A 10 year old boy with CF is noted to have increased bronchovascular markings
48 and new cystic cha...' Yes Is Selected

49 Sc3b Is there a minimum time since his last CT scan that you would want to see elapsed
50 before you repeat the CT scan?

- 51 Yes
52 No
53

54
55 Answer If 'Is there a minimum time since his last CT scan that you would want to see
56 elapsed before you repeat the CT scan?' Yes Is Selected

57 Sc3c What is this minimum time in months?
58
59
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Sc3 Comments

Sc4 A 10 year old girl with CF has deteriorated over a 6 month period. She has increased cough and her FEV1 has fallen from 85% to 70% predicted. There is limited improvement with course of intravenous antibiotics. Her total IgE is 1500 and she has positive specific IgE and precipitins to *Aspergillus fumigatus*. There are no new changes on her CXR. Would you routinely perform a CT scan to aid the diagnosis of ABPA?

- Yes
- No

Sc4 Comments

Sc5 A 10 year old girl with CF develops a left lower lobe collapse during an infective exacerbation. Despite admission for 2 weeks intravenous antibiotics and intensive physiotherapy, the lobe does not re-inflate. She is therefore booked for a flexible bronchoscopy. Would you undertake a CT scan in this girl?

- No - not at any point
- Yes - as soon as the chest x-ray confirmed the left lower lobe collapse
- Yes - but only after two weeks of antibiotics and physiotherapy have failed to re-inflate the lobe
- Yes - but only if bronchoscopy is unsuccessful at re-inflating the lobe

Sc5 Comments

Final Do you have any further comments regarding the use of CT scans in children with cystic fibrosis?

CT Radiologists

Q1 Which centre are you from?

Q2 What is the make and model of the primary CT scanner that is used to scan children at your centre?

- Siemens
- GE
- Toshiba
- Phillips
- Other (please specify) _____

Q3 Model:

Q4 What type of chest CT scans do you perform in children with CF? (Select all applicable)

- Volume / helical scan
- HRCT / axial

Q5 What influences your choice of protocol (i.e. helical vs. axial) e.g. age of patient, patient co-operation etc...?

Q6 When performing chest CT, do you routinely acquire expiratory images in children with CF?

- Yes
- No
- Don't know

Q7 If you do acquire expiratory images (even if non-routine), how is this done?

- Breath holding
- Decubitus positioning
- Other (please specify) _____
- Never acquire
- Don't know

Q8 Do you routinely use contrast when performing chest CT scans in children with CF?

- Yes
- No
- Don't know

Answer If Do you routinely use contrast? No Is Selected

Q8A Are there any circumstances in which you would use contrast to scan the chest of a child with CF? Please give details

1
2
3 Q9 Do you ever scan children with CF under general anaesthetic?

- 4 Yes
5 No
6 Don't know
7

8 Answer If Do you ever scan children with CF under general anaesthetic? Yes Is Selected

9 Q9A Under what circumstances would you use general anaesthetic (e.g. in which age
10 groups)?
11

12
13 Q10 Do you ever use sedation to scan children with CF?

- 14 Yes
15 No
16 Don't know
17

18 Answer If Do you ever use sedation to scan children with CF. Yes Is Selected

19 Q10A Under what circumstances would you use sedation (e.g. in which age groups)?
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22 Q11 Do you have any further comments regarding the use of CT scans in children with CF?
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