BMJ Paediatrics Open

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

Francis J Gilchrist,[1,2] Richard Buka,[2] Mary Jones,[3] Sheng Ang Ho,[2] Warren Lenney,[2] William D Carroll[1,2]

To cite: Gilchrist FJ, Buka R, Jones M, *et al*. Clinical indications and scanning protocols for chest CT in children with cystic fibrosis: a survey of UK tertiary centres. *BMJ Paediatrics Open* 2018;**2**:e000367. doi:10.1136/bmjpo-2018-000367

► Additional material is published online only. To view please visit the journal online (http://dx.doi.org/10.1136/bmjpo-2018-000367).

Received 5 September 2018
Revised 3 October 2018
Accepted 7 October 2018

[1]Institute of Applied Clinical Science, Keele University, Keele, UK
[2]Academic Department of Child Health, Royal Stoke University Hospital, University Hospitals of North Midlands NHS Trust, Stoke on Trent, UK
[3]Department of Radiology, Royal Stoke University Hospital, Stoke on Trent, UK

Correspondence to
Dr Francis J Gilchrist; francis.gilchrist@uhnm.nhs.uk

ABSTRACT

Objectives Chest 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 cooperation 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 already known on this topic?

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

What this study hopes to add?

- There was marked variation among UK centres in the clinical indications for chest CT in children with CF.
- There was marked variation in the protocols used by radiographers when obtaining chest CT scans in children with CF.
- 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] 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 interobserver and intraobserver agreement for the scoring systems, especially in young children.[10] The benefits of chest CT must be balanced against the subsequent lifetime risk of malignancy associated with ionising radiation.[7 11] This is relevant in CF as life expectancy has increased beyond the age at

which such malignancies present.[12] The introduction of modern scanners and the use of paediatric-specific scan protocols has reduced the radiation dose associated with CT scans.[13] Despite this, the cumulative radiation dose for children with CF is substantial, and chest CT is the major contributor.[14 15]

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

AIMS

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

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 antibiotics, with new chest X-ray (CXR) changes, with ABPA and with lobar collapse. This questionnaire can be seen in online supplementary 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.

A separate questionnaire was developed to identify the technical details of chest CT when performed in children with CF. It collected data on the make and model of scanner, the type of scans performed, the use of sedation or general anaesthetic (GA), the use of intravenous contrast and the acquisition of expiratory images. This questionnaire can be seen in online supplementary appendix 1B. An electronic link to this questionnaire was sent to the superintendent radiographer at each of the 27 UK paediatric CF centres.

RESULTS

Clinical indications

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

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 cooperate. 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. GA was used if necessary by 16 (73%) of centres. Indications for GA included the child being unable to cooperate with a breath hold, being of a young age or having learning difficulties. Only 1 (5%) centre reported the routine use of contrast and 3 (14%) routinely obtained expiratory images. When expiratory images were obtained, 14 (64%) used breath-holding command, 5 (23%) relied on ventilation by the anaesthetist during GA and 3 (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 practised 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

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 FEV_1 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 intravenous antibiotics and physiotherapy unsuccessful at reinflating lobe	5	19
	Yes, if bronchoscopy unsuccessful at reinflating lobe	15	56
	Not at any point	6	22

CXR, chest X-ray; FEV_1, forced expiratory volume in 1 s, NTM, non-tuberculous mycobacteria.

guidelines.[16 17] In contrast, there was wide variation in the use of chest CT in other common CF clinical scenarios (reduced forced expiratory volume in 1 s with no response to intravenous antibiotics, new cystic changes on CXR and lobar collapse) for which there is currently no UK guideline. The National Institute for Health and Care Excellence 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] While 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 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 from chest CT alone being approximately 9 mSv if biennial scans have been performed and 18 mSv if annual scans are performed. This compares with 2.8 mSv when chest CT is only performed when clinically indicated.[14] To put these doses into context, the annual background radiation dose in the UK is approximately 2.7 mSv.[21] The radiation dose associated with a CT scan depends on the region of the body being scanned, the type of scan, the age/size of the child and dose saving features used. It is therefore unsurprising that although clinicians reported being aware of the radiation dose associated with chest CT at their centre, the reported dose varied more than 500-fold (0.04–2.1 mSv). This difference is far higher than can be explained by the different protocols. A recent single-centre study reported the estimated effective dose from chest CT in children to be 0.57–2.79 mSv for helical scans and 0.22–0.59 mSv for axial scans.[14]

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 s 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.[22] There is a risk of iatrogenic atelectasis on chest CT performed under GA.[23] 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.[23]

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

CONCLUSIONS

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

Contributors FJG conceptualised and designed the study; he developed the questionnaires and wrote the first draft of the article article. RB assisted with the collection and analysis of data. MJ and WL helped develop the questionnaires. MJ, SAH, WL and WDC assisted with analysis and interpretation of results. All authors reviewed and revised the manuscript and approved the final manuscript as submitted.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Not required.

Provenance and peer review Not commissioned; externally peer reviewed.

Data sharing statement All data from this article is available upon request to the corresponding author.

REFERENCES

1. Elborn JS. Cystic fibrosis. *The Lancet* 2016;388:2519–31.
2. Tiddens HA, Stick SM, Davis S. Multi-modality monitoring of cystic fibrosis lung disease: the role of chest computed tomography. *Paediatr Respir Rev* 2014;15:92–7.
3. Brody AS, Klein JS, Molina PL, *et al*. High-resolution computed tomography in young patients with cystic fibrosis: distribution of abnormalities and correlation with pulmonary function tests. *J Pediatr* 2004;145:32–8.
4. Sly PD, Brennan S, Gangell C, *et al*. Lung disease at diagnosis in infants with cystic fibrosis detected by newborn screening. *Am J Respir Crit Care Med* 2009;180:146–52.
5. Stick SM, Brennan S, Murray C, *et al*. Bronchiectasis in infants and preschool children diagnosed with cystic fibrosis after newborn screening. *J Pediatr* 2009;155:623–8.
6. de Jong PA, Lindblad A, Rubin L, *et al*. Progression of lung disease on computed tomography and pulmonary function tests in children and adults with cystic fibrosis. *Thorax* 2006;61:80–5.
7. Kuo W, Ciet P, Tiddens HA, *et al*. Monitoring cystic fibrosis lung disease by computed tomography. Radiation risk in perspective. *Am J Respir Crit Care Med* 2014;189:1328–36.
8. de Jong PA, Nakano Y, Lequin MH, *et al*. Dose reduction for CT in children with cystic fibrosis: is it feasible to reduce the number of images per scan? *Pediatr Radiol* 2006;36:50–3.
9. Stick S, Tiddens H, Aurora P, *et al*. Early intervention studies in infants and preschool children with cystic fibrosis: are we ready? *Eur Respir J* 2013;42:527–38.
10. Thia LP, Calder A, Stocks J, *et al*. Is chest CT useful in newborn screened infants with cystic fibrosis at 1 year of age? *Thorax* 2014;69:320–7.
11. Pearce MS, Salotti JA, Little MP, *et al*. Radiation exposure from CT scans in childhood and subsequent risk of leukaemia and brain tumours: a retrospective cohort study. *Lancet* 2012;380:499–505.
12. Neglia JP, FitzSimmons SC, Maisonneuve P, *et al*. The risk of cancer among patients with cystic fibrosis. cystic fibrosis and cancer study group. *N Engl J Med* 1995;332:494–9.
13. Lee C, Pearce MS, Salotti JA, *et al*. Reduction in radiation doses from paediatric CT scans in Great Britain. *Br J Radiol* 2016;89:20150305.
14. Ward R, Carroll WD, Cunningham P, *et al*. Radiation dose from common radiological investigations and cumulative exposure in children with cystic fibrosis: an observational study from a single UK centre. *BMJ Open* 2017;7:e017548.
15. de Jong PA, Mayo JR, Golmohammadi K, *et al*. Estimation of cancer mortality associated with repetitive computed tomography scanning. *Am J Respir Crit Care Med* 2006;173:199–203.
16. Thia LP, Balfour Lynn IM. Diagnosing allergic bronchopulmonary aspergillosis in children with cystic fibrosis. *Paediatr Respir Rev* 2009;10:37–42.
17. Floto RA, Olivier KN, Saiman L, *et al*. US Cystic Fibrosis Foundation and European Cystic Fibrosis Society consensus recommendations for the management of non-tuberculous mycobacteria in individuals with cystic fibrosis. *Thorax* 2016;71 Suppl 1:i1–i22.
18. NICE. Cystic fibrosis: diagnosis and management | Guidance and guidelines. https://www.nice.org.uk/guidance/ng78/chapter/recommendations (accessed 27 Sep 2018).
19. Borowitz D, Robinson KA, Rosenfeld M, *et al*. Cystic Fibrosis Foundation evidence-based guidelines for management of infants with cystic fibrosis. *J Pediatr* 2009;155:S73–S93.
20. Lahiri T, Hempstead SE, Brady C, *et al*. Clinical practice guidelines from the cystic fibrosis foundation for preschoolers with cystic fibrosis. *Pediatrics* 2016;137:e20151784.
21. NRPB. *Living with Radiation'*. 5 edn: NRPB, Chilton, Didcot, 1998.
22. Malviya S, Voepel-Lewis T, Eldevik OP, *et al*. Sedation and general anaesthesia in children undergoing MRI and CT: adverse events and outcomes. *Br J Anaesth* 2000;84:743–8.
23. Newman B, Krane EJ, Gawande R, *et al*. Chest CT in children: anesthesia and atelectasis. *Pediatr Radiol* 2014;44:164–72.