Cumulative Radiation Exposure in Children with Cystic Fibrosis

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Introduction

Cystic fibrosis (CF) is the most common autosomal recessive disease in Western Europe with a prevalence of 1 in 1,461 births. It is a multisystem disorder affecting primarily the respiratory and gastrointestinal tracts, caused by mutations in the cystic fibrosis transmembrane conductance regulator protein (CFTR) gene. Aggressive clinical management including proactive treatment of respiratory infections, nutritional support, physiotherapy and newer medication regimens have significantly improved life expectancy. The predicted median survival for babies born in the United Kingdom in the 21st century is now more than 50 years. Death is primarily due to respiratory failure. 

Imaging studies that use ionising radiation are a useful and necessary tool for evaluating and monitoring disease progression and response to treatment in CF. CT scans in particular are associated with high radiation dose. Children are ten times more sensitive to radiation than adults with girls twice as radiosensitive as boys. Children have a longer lifetime over which to develop cancer and developing organs such as the thyroid, bone marrow, breast, brain and skin are especially vulnerable to radiation dose increases with age, up to 15 years. Interestingly those aged 15-20 years had a lower radiation dose than those aged 10-15 years. This may be explained by the fact that the care of all cystic fibrosis patients was transferred at our institution to a paediatric or adult, and provides valuable baseline information against which effects of radiation on the paediatric CF population currently attending our centre.

To our knowledge this is the first study examining cumulative radiation exposure in the Irish CF population, either paediatric or adult, and provides valuable baseline information against which effects of chronic radiation exposure may be measured. As expected, we documented that cumulative radiation exposure increases with age, up to 15 years. Interestingly those aged 15-20 years had a lower mean cumulative radiation dose than those aged 10-15 years. This may be explained by the fact that the care of all cystic fibrosis patients was transferred at our institution to a paediatric or adult, and provides valuable baseline information against which effects of radiation on the paediatric CF population currently attending our centre.

Discussion

This retrospective study calculates an estimate of the average cumulative radiation dose of the paediatric CF population currently attending our centre. To our knowledge this is the first study examining cumulative radiation exposure in the Irish CF population. Data on the number of studies done for all patients, length of follow-up, mean age, mean sex and mean range of age groups were retrospectively reviewed and where sufficient data on exposure factors and technique were available to do so, an estimate of radiation dose was calculated using the ImPACT patient dosimetry calculator and National Radiological Protection Board (UK) data files. 

Results

Data was collected for 77 children, 27 girls and 50 boys, with a mean age of 8.5 (1.9-25.7) years and a follow up time of 686 person years. The average follow up time was 8.5 years. The 77 patients had an average of 19.3 chest radiographs each. Thirty children had an average of 4.7 abdominal radiographs each; 26 children had an average of 1.1 barium meals each; 11 patients had an average of 2.1 enemas each and 29 patients had 1.7 C.T. scans each.

There was no significant correlation between lung function and cumulative radiation exposure. No association was identified between microbiology cultures and type of radiological studies performed; and the number of other x-ray or contrast examinations done (p=0.0004). Radiation dose increased with patient age (r=0.36, p=0.0014) (see Figure 1).

The average cumulative radiation dose was 6.2 (0.04-25) mSv per C.F. patient. There was no significant difference in cumulative radiation dose between boys and girls (p=0.47). The average cumulative radiation dose was greater in children who presented with meconium ileus. Admission of the cumulative radiation dose was greater in children who presented with meconium ileus (n=12), compared with only 2 contrast enemas in children without meconium ileus. When the radiation dose of these enemas was subtracted, there was no significant difference in cumulative radiation dose between boys and girls (p=0.47).

Fifteen percent (n=12), presented with meconium ileus in the newborn period. Cumulative radiation dose was greater in these patients (4.5mSv), compared with those who did not present with meconium ileus (n=65), (5.5mSv), (p=0.46). The cumulative radiation dose in each age group is shown in Table 1. Cumulative radiation dose increased with patient age (r=0.36, p=0.0014) (see Figure 1).

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In our paediatric CF population who were old enough to perform reproducible lung function testing, there was no correlation between lung function and cumulative radiation dose. Traditionally, disease progression in CF has been monitored by a combination of clinical assessment, chest radiographs and pulmonary function tests. Increase in Brasfield FEV1 clinical assessment, chest radiographs and pulmonary function tests. Increase in Brasfield FEV1 in our paediatric CF population who were old enough to perform reproducible lung function (n=44, 71%). An adult CF population with more advanced lung disease and more severe reduction in lung function tests may demonstrate an increased cumulative radiation dose. FEV1 is employed internationally as a marker for severity of respiratory involvement in CF and a predictor of death, though its limitations are well documented. Performance of lung function tests even Uhthoff children and older children with cystic fibrosis, is technically much more challenging in infants and preschool children. Lung function tests are an indirect measure of lung structure and are insensitive to localised or early damage in pulmonary function may be preceded by structural change detected on CT scan young patient, in the absence of lung function tests. CT scans, where clinically indicated, may be very useful in identifying disease progression.

Analysis of patient microbiology cultures (n=77) in particular Staph aureus (n=44) and Pseudomonas aeruginosa (n=29) did not reveal any correlation with cumulative radiation dose. For patients with chronic Pseudomonas aeruginosa on cough swab culture and sensitivity. Chronic infection with P. aeruginosa which causes a more rapid decline in lung function than Staph aureus, is an important predictor of survival and is the most important cause of mortality. Expect that people with CF who are chronically infected with P. aeruginosa as opposed to Staph aureus would have more reduced lung function and an associated higher cumulative radiation dose. Our study did not demonstrate a correlation between microbiology cultures and cumulative radiation dose. This may be explained by our relatively small number of patients infected with P. aeruginosa and a relatively small percentage of patients with clinically confirmed P. aeruginosa infection. We documented that cumulative radiation dose increases with the number of CTs performed. Providing a UK study documented that while a small number of CTs was associated only 4% of radiological procedures in the UK, CT contributed to 40% of the population radiation dose from medical exposures. CT is also the largest contributor to cumulative radiation dose in our study.

The radiation dose per CT in CF children can vary widely depending on the technique used. While one study estimated the mean radiation dose per CT to be 6.6mSv (range 1.5-29.3) others estimated it to be much less than 1mSv per CT. CF patients with annual CT thorax, assuming median survival to 36 years, risks are likely flawed because they are calculated from effects of radiation exposure on survivors of the Japanese atomic bombs. An observational study would require follow up of hundreds of thousands of people for their entire lifetime to have sufficient power to detect cancer risk. No data exists regarding the radiation risk with relatively small doses associated with medical imaging. Although irising radiation dose per CT study has reduced significantly over the past 20 years, this may be partially counterbalanced by both the increased use of CT of CF and the increased median survival of this population.

Cumulative radiation exposure in our paediatric CF population compares favourably with that in other institutions worldwide where routine CT thorax is performed. Cumulative radiation dose increases with age, number of CT scans, and at our institution, a history of meconium ileus. In our group, no association was identified between cumulative radiation dose and microbiology cultures or lung function as measured by FEV1. We recommend that physicians and radiologists work together discussing each individual case, aiming for a scan that maximises disease identification while minimising radiation exposure, as is done in our institution. newer scanners, breast shields, reduction of tube kV and mA and current all reduce radiation dose in a susceptible population based on young age and life long radiation exposure.

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