LUNG FUNCTION AND COMPUTED TOMOGRAPHY (CT) FINDINGS IN INFANTS WITH CYSTIC FIBROSIS DIAGNOSED BY NEWBORN SCREENING

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**Aim:** To measure lung function in infants with cystic fibrosis (CF) diagnosed by newborn screening, and correlate findings with lung structure.

**Methods:** Prospective recruitment of infants and young children less than 2.5 years of age diagnosed with CF by newborn screening. Subjects had infant lung function measured and CT scan performed shortly after diagnosis, and/or around the time of their birthday. Lung function was measured using the raised volume rapid thoraco-abdominal compression technique at an inflation pressure of 20 cmH2O. A cohort of health control infants who were drawn from the general population had infant lung function testing performed under identical testing conditions. From our control cohort we used multiple linear regression to develop a prediction equation, which allowed us to express the FEV0.5 data as z-scores. CT was performed under general anaesthetic. Limited axial scans were obtained above the carina, below the carina and above the diaphragm using 1.5mm collimations at 20cmH2O and repeated at passive exhalation. The images were then scored using a modification of the Brody score.

**Results:** We tested 71 infants (on 90 occasions) with CF and 49 controls aged from 5 weeks to 2.2 years. The z-score for FEV0.5 measured shortly after the diagnosis of CF by newborn screening was 0.03 (95% CI -0.38 to 0.44, p=0.88), indicating little evidence that the z-score at first measurement differed from zero, in other words it was normal. The z-score was diminished by 0.77 for each year older infants were at time of testing (95% CI -1.14 to -0.41, p<0.001) indicating strong evidence that the decrease is greater than zero. This occurred even though the infants had been diagnosed by newborn screening, were cared for in a specialist centre and remained well nourished throughout. This is the first time this has been shown. 81 CT scans were scored. Modified Brody score ranged from 0 (no abnormalities) to19/48. The total score in this cohort was primarily composed of the air-trapping and bronchial wall thickening components. Bronchiectasis, parenchymal changes, and bronchiolitis were rarely detected. There was no association demonstrated between diminished lung function and evidence of structural changes as detected by the total modified Brody score or the sub-components of the total score.

**Conclusions:** FEV0.5 is within the normal range when tested shortly after the diagnosis of CF by newborn screening, but is diminished in older infants despite good nutrition and ongoing care in a specialist centre. The diminished FEV0.5 is independent of structural changes as detected by a modified Brody score of a limited slice CT.