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Title: Normal lung function in infants with cystic fibrosis shortly after birth

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Body: Background In children with cystic fibrosis (CF) small airway disease can occur early in life. Objective To assess ventilation heterogeneity, a marker of small-airway function, shortly after birth in infants with CF diagnosed by newborn screening. Methods We performed multiple-breath washout (MBW) using 4% sulfur hexafluoride and tidal breathing measurements in 23 infants with CF, aged median (range) 6.3 (3.9 – 12.6) weeks and compared it to a previously reported equipment and tracer-gas specific normative data population of 305 healthy infants, aged median 5.0 (3.6 – 8.7) weeks (Fuchs et al, ERJ 2011). We compared lung clearance index (LCI), functional residual capacity (FRC) and the following tidal breathing parameters: Tidal volume, respiratory rate, minute ventilation, mean and peak tidal inspiratory and expiratory flow and the ratio of time to peak tidal expiratory flow to expiratory time. Results Compared to controls, and after adjustment for body weight and age, LCI was normal in infants with CF (mean difference (95% CI) 0.28 (-0.19 to 0.74) z-scores, p = 0.24), as was FRC (mean difference (95% CI) 0.40 (-0.05 to 0.85) z-scores, p = 0.08) and tidal breathing parameters. Only one infant with CF had elevated LCI (> 1.96 z-scores), while none showed elevated FRC. Conclusions CF infants show normal LCI values shortly after birth and thus no sign of ventilation inhomogeneity. The fact that all other parameters are also normal reflects the still undamaged state of the lungs. This highlights the importance of early therapy to maintain normal lung function as long as possible.