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Title: Stable lung function is maintained over 2 years in newborn screened (NBS) CF infants

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Body: Background:Lung function in LCFC NBS CF infants was impaired by age 3-months (3m) compared to healthy controls.[1]However forced expiratory volume ($FEV_{0.5}$) improved by 1-year (1y) while lung clearance index (LCI) and plethysmographic functional residual capacity (FRC) remained stable.[2] These findings contrast with previous reports of progressive deterioration of lung function in NBS CF infants. Hypothesis:In NBS CF infants, lung function is stable to 2y. Methods:LCI, FRC and $FEV_{0.5}$ were measured in NBS CF infants and controls at 3m, 1y and 2y. Results:To date, 50 CF and 24 control infants have completed 3 tests. Mildly elevated LCI and FRC remained stable from 3m-1y with no further deterioration by 2y. LCI and FRC were on average higher in CF whereas a significant reduction in $FEV_{0.5}$ was only evident at 3m. Longitudinal changes in LCI and FRC were similar in both groups; whilst significant improvement in $FEV_{0.5}$ for CF from 3m-1y was significantly different from the lack of change in controls.

Lung function in CF infants

	Mean (SD)			Mean (95% CI) change	
	3m	1y	2y	1y-3m	2y-1y
Z-$FEV_{0.5}$	-1.4 (1.1)	-0.4 (1.1)	-0.4 (1.0)	1.0 (0.7;1.3)¤	0.0 (-0.3;0.3)

Z-LCI	0.7 (1.3)	1.0 (1.2)	0.9 (1.0)	0.3 (-0.1;0.7)	-0.0 (-0.4;0.4)
Z-FRC	1.0 (1.0)	0.9 (1.0)	1.0 (1.5)	-0.2 (-0.5;0.2)	0.2 (-0.2;0.5)

Healthy controls not shown. Results in bold = signif diff ($p<0.05$) CF-controls. ¤Signif diff in $FEV_{0.5}$: CF 1y-3m ($p<0.001$)

Conclusions:This is the 1st study to demonstrate stable lung function to 2y in NBS CF infants managed on standard CF therapy. These results suggest that in these infants, novel treatments could be deferred beyond infancy when objective outcomes are more easily measured. References:[1]Hoo et al. Thorax 2012;[2]Thia et al. ERJ 2011(S55).