

Online Data Supplement

Heritability of Lung Disease Severity in Cystic Fibrosis

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Table E1: Intra-pair Correlations for Cross-sectional and Longitudinal Measures of CF Lung Disease Severity for Twins and Siblings Less than 15 Years of Age and Greater than 15 Years of Age, Living Together

	MaxFEV1CF%	AvgFEV1CF%	EstFEV1%@20yrs
MZ Twins, Both <15 Years of Age	0.90 (14) [*]	0.94 (8) ^{**}	0.84 (7) ^{***}
MZ Twins, Both >15 Years of Age	0.88 (22) [*]	0.89 (25) [*]	0.85 (24) [*]
Same Sex DZ Twins and Siblings, Both <15 Years of Age	0.55 (46) ^{**}	0.67 (31) [*]	0.51 (24) ^{***}
Same Sex DZ Twins and Siblings, Both >15 Years of Age	0.53 (16) ^{***}	0.61 (16) ^{***}	0.54 (15) ^{***}

* p-value < 0.0001

** p-value < 0.001

*** p-value < 0.05

Table E2: Correlations for Cross-Sectional and Longitudinal Measures of CF Lung Disease Severity in Δ F508 Homozygous Twins and Siblings Living Together at Home

	Max- FEV1CF% (n)	EstFEV1% @20yrs[†] (n)	Adjusted¹ EstFEV1% @20yrs (n)	AvgFEV1- CF%^{††} (n)	Adjusted² AvgFEV1- CF% (n)
Monozygous Twins	0.92 ± 0.01 (23) [*]	0.86 ± 0.02 (22) [*]	0.82 ± 0.02 (22) [*]	0.93 ± 0.01 (22) [*]	0.90 ± 0.01 (20) [*]
Dizygous Twins	0.59 ± 0.13 (4)	-0.18 ± 0.60 (3)	0.61 ± 0.24 (3)	0.40 ± 0.37 (3)	0.75 ± 0.10 (3)
Siblings	0.28 ± 0.01 (93) ^{****}	0.36 ± 0.02 (48)	0.46 ± 0.02 (48) ^{****}	0.41 ± 0.01 (67) ^{**}	0.38 ± 0.01 (64) ^{***}
Same Sex Siblings, <3 years difference in age	0.50 ± 0.02 (31) ^{****}	0.59 ± 0.03 (21) ^{****}	0.45 ± 0.04 (21)	0.54 ± 0.02 (26) ^{***}	0.51 ± 0.02 (25) ^{**}
Same Sex DZ Twins and Siblings, < 3 years difference in age	0.49 ± 0.02 (34) ^{****}	0.58 ± 0.03 (23) ^{****}	0.44 ± 0.03 (23)	0.53 ± 0.02 (28) ^{***}	0.51 ± 0.02 (27) ^{****}

¹ EstFEV1%@20yrs adjusted for AvgBMIZ and for pancreatic status

² AvgFEV1CF% adjusted for AvgBMIZ

[†] Using minimum of 5 years of PFT data

^{††} Using minimum of 4 years of PFT data

^{*} p-value < 0.0001

^{**} p-value < 0.001

*** p-value < 0.005

**** p-value < 0.01