

Online Supplement

Quality of Life and Mortality Outcomes for Augmentation Naïve and Augmented Patients with Severe Alpha-1 Antitrypsin Deficiency

Paul R. Ellis, MBChB, PhD¹ Kristen E. Holm, PhD, MPH^{2,3} Radmila Choate, PhD, MPH⁴ David M. Mannino, MD⁴ Robert A. Stockley, MD, DSc⁵ Robert A. Sandhaus, MD^{3,6} Alice M. Turner, MBChB, PhD¹

¹Institute of Applied Health Research, University of Birmingham, Birmingham, United Kingdom

²Division of Neurology and Behavioral Health, National Jewish Health, Denver, Colorado, United States

³AlphaNet, Kissimmee, Florida, United States⁴University of Kentucky College of Public Health, Lexington, Kentucky, United States

⁵Lung Investigation Unit, Queen Elizabeth Hospital Birmingham, University Hospitals Birmingham, National Health Service Foundation Trust, Birmingham, United Kingdom

⁶Division of Pulmonary, Critical Care and Sleep Medicine, National Jewish Health, Denver, Colorado, United States

Supplementary figures

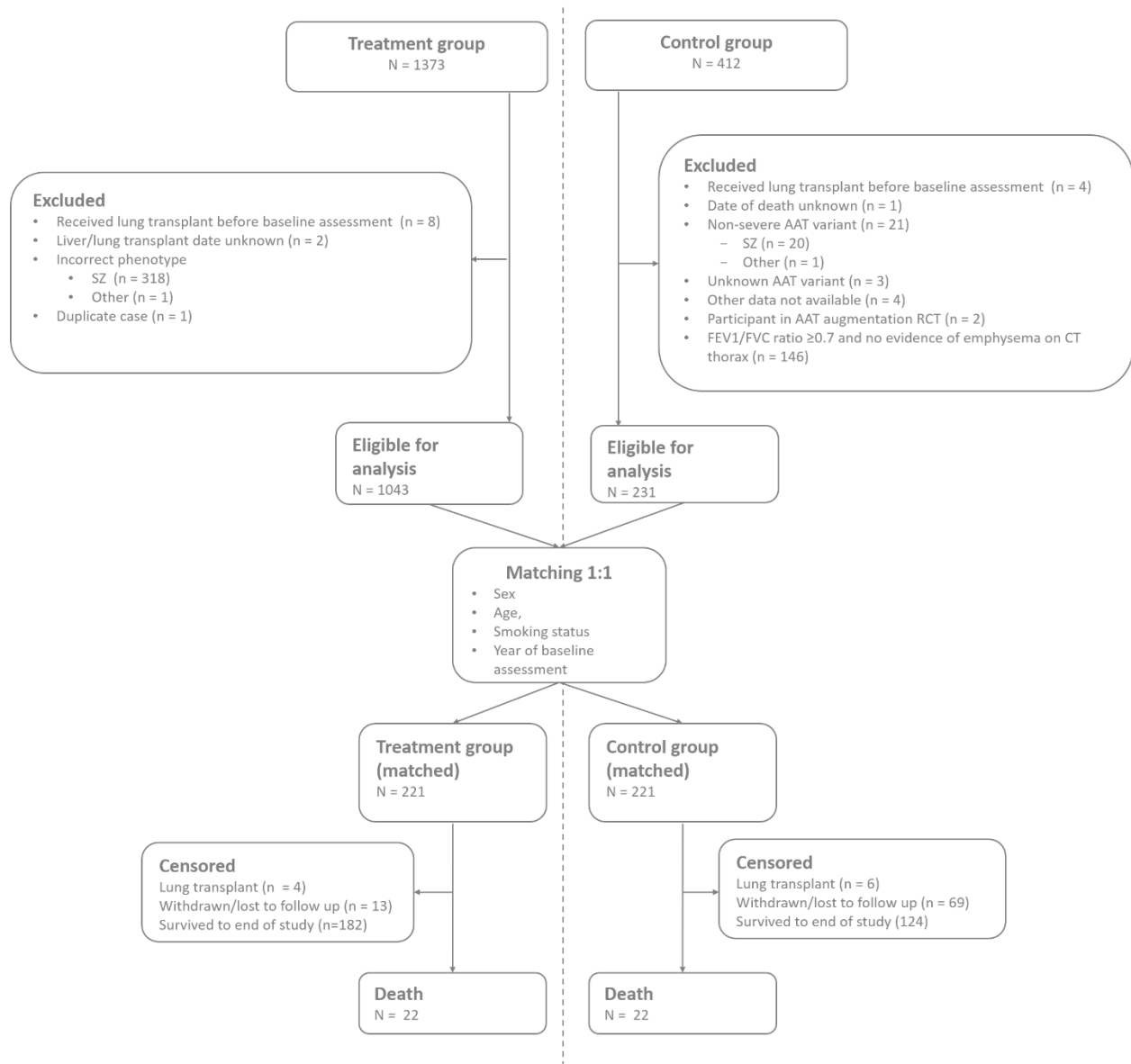


Figure S1: Study flow diagram for the whole cohort.

Supplementary tables

Table S1: Univariate and multivariate analysis for annual change in SGRQ total and symptom scores

	Estimate	95% CI	p
Univariate Analysis			
Change in SGRQ total/year			
Sex (Female)*	0.65	-0.12 to +1.41	0.098
Age*	0.03	-0.00 to +0.07	0.087
Smoking status	-0.54	-1.39 to +0.30	0.21
(Ex-smoker)			
Smoking status	-0.80	-2.91 to +1.31	0.46
(Current smoker)			
Control group*	1.19	0.26 to 2.12	0.013
FEV ₁ % predicted	-0.01	-0.03 to +0.01	0.28
Baseline SGRQ total*	-0.07	-0.09 to -0.05	<0.001
Change in SGRQ⁺ symptom/year			
Female	0.46	-0.66 to +1.58	0.42
Age	0.04	-0.02 to +0.09	0.17

Smoking status	-0.04	-1.28 to +1.21	0.95
(Ex-smoker)			
Smoking status (Current smoker)	-0.99	-4.08 to +2.10	0.53
Control Group*	1.21	-0.15 to +2.58	0.081
FEV ₁ % predicted	-0.00	-0.03 to +0.02	0.78
Baseline SGRQ symptom*	-0.10	-0.12 to -0.07	<0.001

Multivariate analysis

Change in SGRQ total/year

Control	1.30	0.41 to +2.19	0.004
Age (years)	-0.01	-0.05 to +0.03	0.65
Female	0.45	-0.29 to +1.19	0.23
Baseline SGRQ total	-0.07	-0.09 to -0.05	<0.001

Change in SGRQ symptom/year

Control Group	2.01	0.73 to 3.30	0.002
Baseline SGRQ symptom	-0.10	-0.13 to +0.08	<0.001

*Variables chosen to be included in multivariate model.

†St. Georges Respiratory Questionnaire (SGRQ) total score is determined by the SGRQ-COPD questionnaire; scores range from 0 to 100 points, with higher scores indicating worse health status (minimum clinically important difference, 4-points(1). SGRQ total score is calculated from three separate health status domains; symptom, impact and activity scores.

SGRQ; St Georges Respiratory Questionnaire, CI; confidence interval, FEV₁; Forced Expiratory Volume in 1 second.