

**Table S1.** Demographic and clinical characteristics in AATD and control children

Variable	Control (n = 38)	Low risk (n = 15)	Intermediate risk (n = 28)	High risk (n = 8)	p-value
<b>Age (median; range)</b>	9 (4-12)	11 (4-14)	9 (4-12)	9 (6-14)	0.182
<b>Gender M/F (n, %)</b>	25/17, 60/40	11/4, 73/27	17/11, 61/39	6/3, 67/33	0.691
<b>AAT (mg/dl)</b>	139.4 (161.0-119.1)	119.5 (127.5-99.0)	82.0 (95.0-67.8)	25.4 (34.2-22.0)	<b>&lt; 0.001</b>
<b>BMI (Kg/m<sup>2</sup>)</b>	18.22 ± 3.77	19.21 ± 2.55	19.17 ± 3.45	19.50 ± 3.31	0.536
<b>FEV<sub>1</sub> (%)</b>	91.90 (88.0-105.4)	100.2 (91.45-109.3)	103.9 (96.20-120.4)	101.0 (100.8-116.0)	0.620
<b>FVC (%)</b>	100.0 (89.8-108.1)	100.0 (89.90-114.5)	108.4 (87.6-119.0)	107.0 (106.9-108.0)	0.951
<b>FEV<sub>1</sub>/FVC (%)</b>	92.0 (78.0-102.0)	85.0 (76.4-107.7)	99.40 (88.8-109.7)	97.2 (83.0-111)	0.432
<b>FEF<sub>25-75</sub> (%)</b>	71.8 (56.7-90.2)	80.5 (59.1-97.3)	104.7 (98.6-108.3)	96.5 (84.1-97.0)	0.095
<b>AST (U/L)</b>	28.00 (24.50-37.50)	26.00 (22.75-30.75)	28.00 (25.00-37.00)	26.00 (25.00-29.00)	0.389
<b>ALT (U/L)</b>	19.00 (18.00-23.50)	18.00 (16.00-20.00)	20.50 (17.25-25.00)	24.00 (19.00-32.00)	0.113
<b>GGT (U/L)</b>	14.00 (12.00-17.25)	15.00 (11.75-16.25)	16.00 (14.00-18.00)	18.00 (17.00-20.00)	0.055

AAT, Alpha-1 antitrypsin; BMI, body mass index; AST, aspartate aminotransferase; ALT, alanine aminotransferase; GGT, gammaglutamyl-transferase; FEV<sub>1</sub>, forced expiratory volume in 1 s; FVC, forced vital capacity; FEF<sub>25-75</sub>, forced expiratory flow 25–75%. Data are expressed as median and interquartile range (IQR). Statistic significance after application of Kruskal-Wallis test. Comparison of proportions was performed by chi-square test. P-values lower than 0.05 were statistically significant (labelled in bold).