

## **Online Supplement**

### **Patient-Reported Pulmonary Symptoms, Exacerbations, and Management in a Cohort of Patients With Alpha-1 Antitrypsin Deficiency**

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**Supplemental Table 1.** Patient-level analysis of the baseline characteristics comparing participants reporting a change in respiratory symptoms in one year and those not reporting changes, n=459 participants

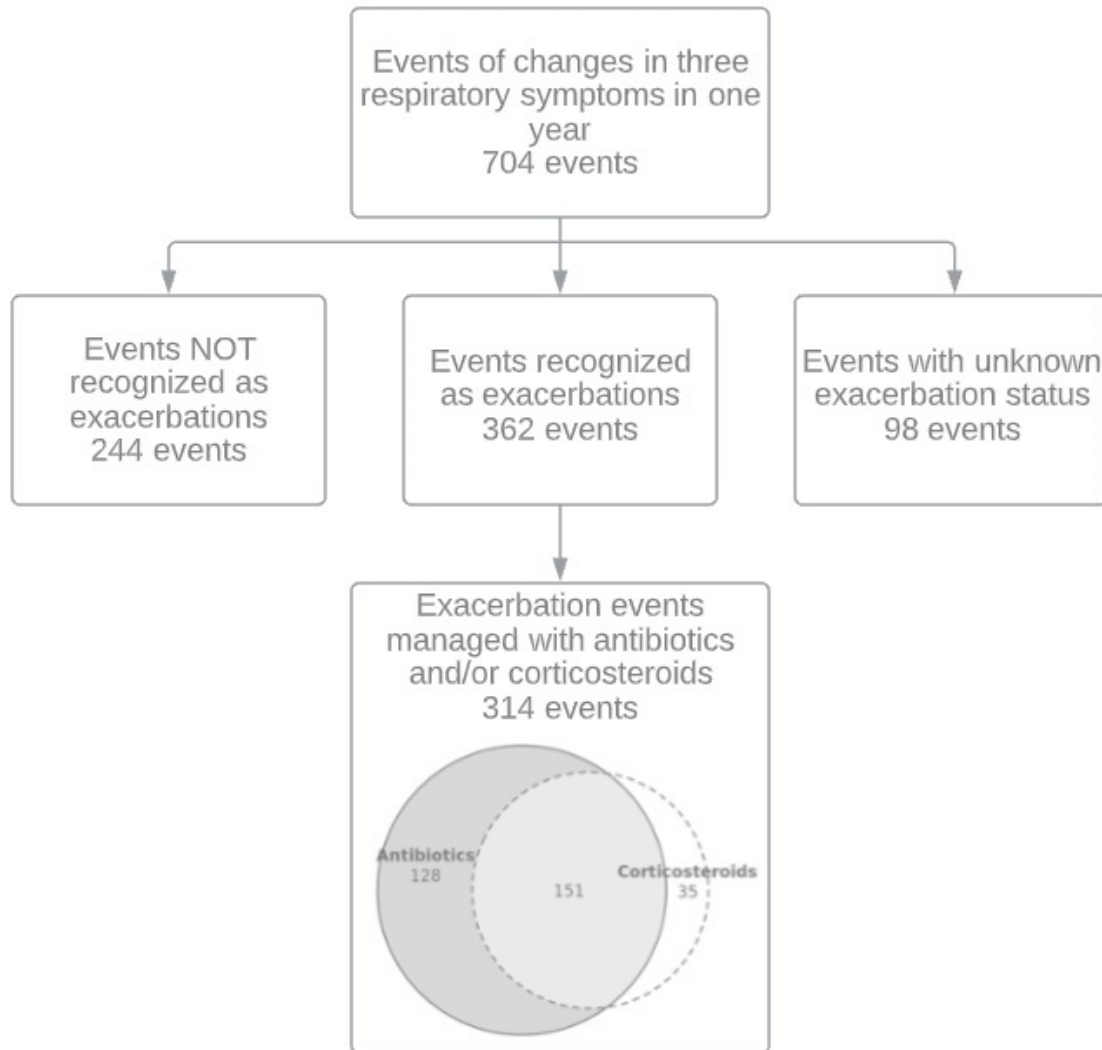
	<b>Participants WITH changes in pulmonary symptoms (n=316)</b>	<b>Participants WITHOUT changes in pulmonary symptoms (n=143)</b>
<b>Age<sup>a</sup>, years, mean (SD)</b>	58.2 (9.7)	57.4 (8.5)
Median	59	57
<b>Female, n (%)</b>	167 (52.9)	78 (55.0)
<b>Married/Civil Union, n (%)</b>	209 (66.8)	112 (78.9)
<b>BMI, mean (SD)</b>	26.06 (5.6)	26.6 (5.2)
Median	25	26
<b>BMI categories, n (%)</b>		
Low (<19)	18 (5.7)	5 (3.5)
Normal (19-25)	137 (43.4)	59 (41.3)
High (>25-30)	102 (32.3)	48 (33.6)
Very High (>30)	59 (18.7)	31 (21.7)
<b>Genotype, n (%)</b>		
ZZ <sup>b</sup>	256 (89.5)	121 (91.7)
SZ	17 (5.9)	6 (4.5)
MZ/Other	13 (4.6)	5 (3.8)
	<i>data available n=286</i>	<i>data available=132</i>
<b>Frequency of exacerbations in the year before baseline, n (%)</b>		
Never	49 (16.0)	46 (35.1)
Once	64 (20.9)	42 (32.1)
Every 6 months	50 (16.3)	17 (13.0)
Every 4 months	41 (13.4)	7 (5.3)
Every 3 months	56 (18.3)	12 (9.2)
Every month	46 (15.0)	7 (5.3)
	<i>data available n=306</i>	<i>data available n=131</i>

<b>SFS Randomization arm, n (%)</b>		
Standard care	159 (50.3)	69 (48.3)
Intervention	157 (49.7)	74 (51.7)
<b>Oxygen use, n (%)</b>	175 (57.2)	49 (37.7)
	<i>data available =306</i>	<i>data available n=130</i>
<b>Self-reported bronchiectasis at baseline, n (%)</b>	48 (15.4)	15 (11.6)
	<i>data available n=311</i>	<i>data available n=129</i>
<b>Ever smoker, n(%)</b>	237 (76.0)	98 (71.0)
	<i>data available n=312</i>	<i>data available n=138</i>
<b>Pack-years, mean (SD)</b>	29.2 (23.8)	25.7 (17.9)
Median	20	20
	<i>data available n=223</i>	<i>data available=96</i>

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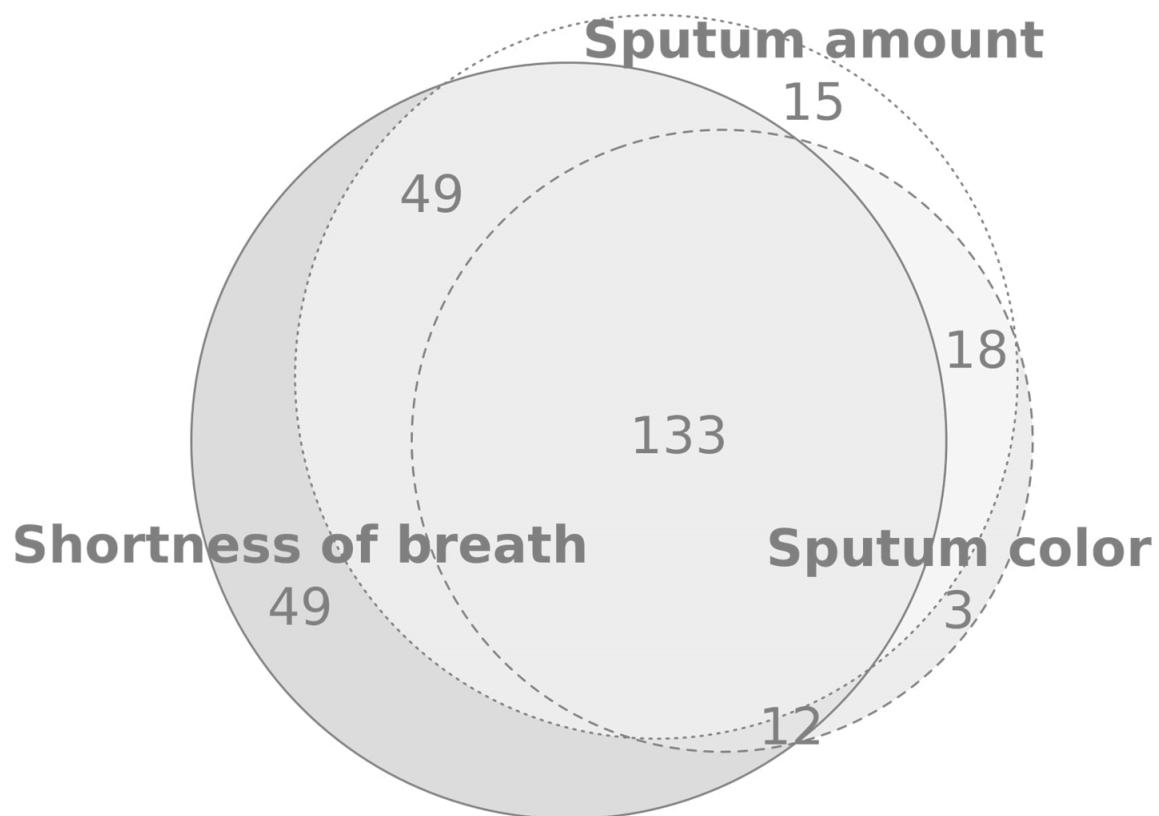
<sup>a</sup> Age calculated as of the first date of the monthly assessment; <sup>b</sup> includes ZNull/NullNull;

**Supplemental Figure 1:**



**Supplemental Figure 1.** Consort diagram of the events of changes in three respiratory symptoms: shortness of breath, change in sputum color and amount, and their management with antibiotics and/or corticosteroids

**Supplemental Figure 2:**



**Supplemental Figure 2.** Venn diagram of *antibiotic treatment* across three respiratory symptoms: shortness of breath, change in sputum color, and change in sputum amount, n=279