

Management of Pulmonary Exacerbations in a Cohort of Patients with Alpha-1 Antitrypsin Deficiency

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Introduction

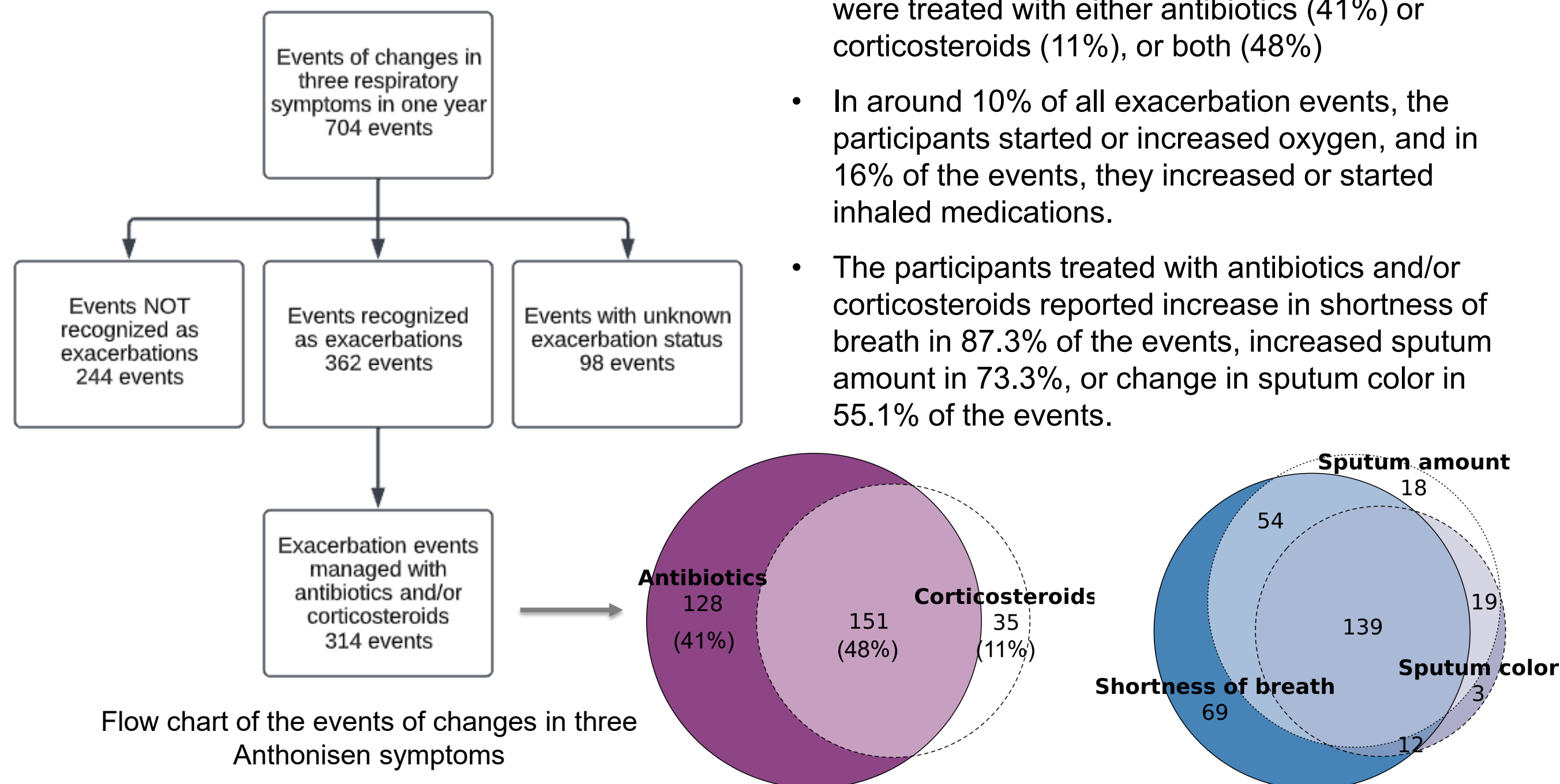
- Exacerbations of alpha-1 antitrypsin deficiency (AATD)-related lung disease present as worsening of respiratory symptoms that often result in additional therapy.
- Effective management of pulmonary exacerbations is essential in reducing their negative impact on disease progression and quality of life.

Methods

- This study used data collected via monthly telephone calls between October 2009 and October 2010 in a cohort of AlphaNet subscribers who participated in AlphaNet's Step Forward Study¹.
- Participants were asked if they experienced changes in their regular respiratory symptoms and whether they considered such changes to be an exacerbation.
- Changes in three respiratory symptoms – increase in shortness of breath, change in sputum color and its amount – were examined considering the clinical significance of these symptoms in identifying exacerbations and guiding management by Anthonisen criteria².
- For the purposes of these analyses, a positive response to changes in one or more of the three Anthonisen symptoms was defined as an "event."

Methods (continued)

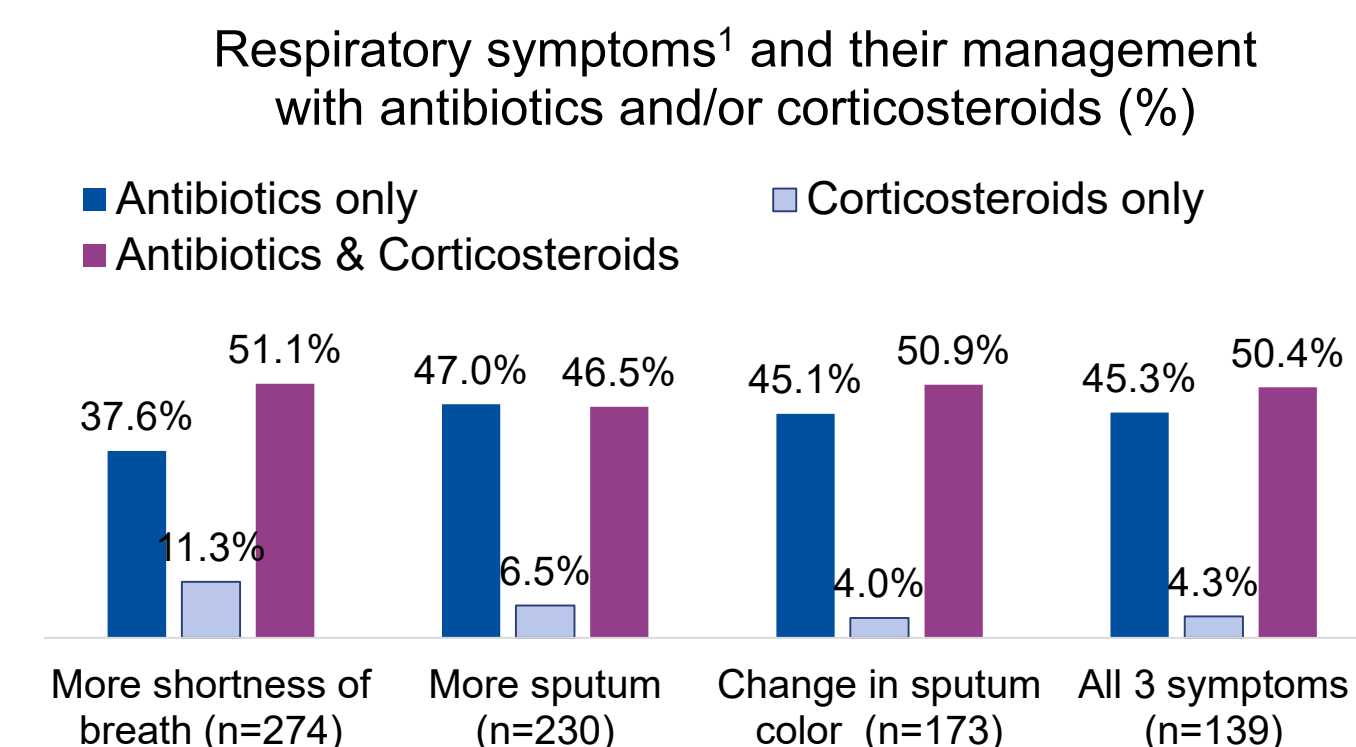
- Participants who identified having an exacerbation were asked about its management.
- Data on exacerbation management using corticosteroids, antibiotics, inhaled medications or use of oxygen were examined. Per-patient and per-event analyses were performed.
- Analyses included participants who reported at least one exacerbation in which there was a change in shortness of breath, sputum color, or sputum amount.



Results

- During the 1-year follow-up, between October 2009 and 2010, 208 SFS participants (mean age 57 ± 10 years, 55% female, 64% married, 79% ZZ variant) reported changes in any of the three Anthonisen symptoms, recognized these changes as exacerbations and were included in this study.
- Overall, 704 events of changes in any of the three respiratory symptoms were reported by the study participants. Over 50% of the events (362/704) were recognized as pulmonary exacerbations.
- Around 87% of the exacerbation events (314/362) were treated with either antibiotics (41%) or corticosteroids (11%), or both (48%)
- In around 10% of all exacerbation events, the participants started or increased oxygen, and in 16% of the events, they increased or started inhaled medications.
- The participants treated with antibiotics and/or corticosteroids reported increase in shortness of breath in 87.3% of the events, increased sputum amount in 73.3%, or change in sputum color in 55.1% of the events.

Results (continued)



¹regardless of the overlap across the three symptoms

Conclusions

- Some AATD-related lung disease exacerbations are managed with conventional COPD treatments, including antibiotics and/or corticosteroids.
- Future research is needed to investigate the effect of AATD-specific management on exacerbation prevention and treatment.

References

- Choate, R, et al. Home-Based Multicomponent Intervention Increases Exercise Activity and Improves Body Mass Index: Results of a 5-Year Randomized Trial Among Individuals with Alpha-1 Antitrypsin Deficiency-Associated Lung Disease. JCO PDF
- Anthonisen, NR et al. Antibiotic therapy in exacerbations of chronic obstructive pulmonary disease. Annals of internal medicine.

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