8.09 Exploring exacerbations and current health trends in severe Alpha-1 antitrypsin deficiency (AATD)

Rosie Carroll^{1,2}, Suzanne Roche^{1,3}, Malcolm Herron^{1,3}, Tomás Carroll^{3,2}, Michelle Casey^{1,3}, Cedric Gunaratnam^{1,3}, Noel. G McElvaney^{1,3}

¹National Centre for Expertise for AATD, Beaumont Hospital, Dublin, Ireland. ²Alpha-1 Foundation Ireland, Dublin, Ireland. ³Irish Centre for Genetic Lung Disease, Royal College of Surgeons in Ireland, Dublin, Ireland

Background: AATD and particularly the severe ZZ phenotype is associated with obstructive lung disease and an increased risk of exacerbations. Exacerbations of AATD are suspected to differ from usual COPD, but precisely how is not clear. Exacerbations are classified based on patient reported change in symptoms or definitive events. We evaluated the frequency and severity of exacerbations in this cohort and explore characteristics of frequent exacerbators. **Methods**: This is a prospective national registry study of ZZ individuals with an FEV1/FVC ratio < 0.7 and/or emphysema on CT. A baseline survey was performed to establish baseline clinical information. Monthly surveys will establish any change in symptoms which could be suggestive of an exacerbation. **Results**: 50 patients are enrolled to date and will be followed for 12 months – data collection is ongoing. Median FEV1 % is 53% and DLCO 50%. A large proportion of patients had combined emphysema and bronchiectasis which together are associated with increased exacerbations. We note a high symptom burden with daily dyspnoea reported in 100% of patients. Early monthly data suggests a large burden of increased symptoms suggesting under-reported exacerbations. **Conclusion:** Exacerbations are frequent with a prolonged recovery and detrimental impact on quality of life. **Conflict of Interest:** The authors declare that they have no conflict of interest.