ETIOLOGY OF NON-CYSTIC FIBROSIS BRONCHIECTASIS IN CROATIA: DATA FROM THE EUROPEAN MULTICENTER BRONCHIECTASIS AUDIT AND RESEARCH COLLABORATION (EMBARC) REGISTRY

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Objective:

Background and objective: Bronchiectasis is one of the most neglected respiratory diseases. The European Multicenter Bronchiectasis Audit and Research Collaboration (EMBARC) registry was established to address the underinvestment in bronchiectasis research, education and clinical care in Europe. We aimed to describe the etiology of non-cystic

fibrosis (non-CF) bronchiectasis in Croatian patients from University Hospital Centre Zagreb (UHC Zagreb) enrolled in the EMBARC registry and compare the data with other published cohorts.

Patients and methods: All patients from UHC Zagreb enrolled in the EMBARC registry were included in the study. The etiology of bronchiectasis was determined after review of the medical history and evidence of testing for allergic bronchopulmonary aspergillosis (ABPA), cystic fibrosis (CF), serum immunoglobulins, α-1 antitrypsin deficiency (A1ATD), serum electrophoresis, test of ciliary function, bronchoscopy and autoantibody testing. CF patients were excluded from the registry.

Results: Among the 65 patients enrolled in the EMBARC registry 33 were female and 32 male, with a mean age of 58 years. The most common cause of non-CF bronchiectasis was post infectious (22/65, 33.84%), followed by COPD (10/65, 15.35%), asthma (7/65, 10.76%) and post tuberculous (6/65, 9.23%). Only 4 patients had idiopathic bronchiectasis (6.15%). 3 patients were diagnosed with Kartagener syndrome (4.6%) and common variable immunodeficiency (CVID) (4.6%), 2 patients with non tuberculous mycobacteria (NTM) (3.07%) while ABPA, primary ciliary dyskinesia, Williams Campbell syndrome, Mounier-Kuhn syndrome, Swyer-James-Macleod syndrome and A1ATD are the cause of bronchiectasis in 1 patient each.

Conclusion: To our knowledge, this is the first description of the etiology of bronchiectasis in Croatian patients. Interestingly, only 6.15% of patients were classified as idiopathic which is much lower than expected in comparison with other European cohorts. Rare causes of

bronchiectasis such as Kartagener syndrome were more common than expected in our cohort. The distribution of other etiologies is in accordance with previously published data. The lower prevalence of idiopathic bronchiectasis could be explained by the small sample size and selection bias, as all the patients are recruited from a single tertiary center in Croatia with greater diagnostic capability in comparison with smaller centers.